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GENERAL OR LOCALIZED HYPOTONIA OF THE MUSCLES IN CHILDHOOD (MYATONIA CONGENITA).

Report of a Case, with Necropsy.

By William G. Spiller, M.D.,
Professor of Neuropathology and Associate Professor of Neurology, University of Pennsylvania.

(From the Neuropathological Laboratory of the University of Pennsylvania.)

All knowledge that we have hitherto obtained concerning congenital atony of the muscles in childhood we owe to Oppenheim. In the September, 1900, number of the *Monatsschrift für Psychiatrie und Neurologie,* vol. viii., Oppenheim published a brief paper concerning this condition. He said that he had had the opportunity repeatedly, especially in recent years, to observe in children in the first months of life—*i. e.,* within the first or second year, this disease condition, concerning which he had not been able to find anything in the literature. The parents complain that the whole body of the child or only certain portions—*i. e.,* the limbs, and especially the lower—are affected; that they are flaccid and motionless. Objectively, hypotonia or even atonia of the muscles is the most striking feature, and this is associated with weakness or loss of the tendon reflexes. The flaccidity is so great that the joints are abnormally movable.

Active motion is always impaired, but in different intensity in the different cases. Where the disorder is most marked the extremities are motionless, and the paralysis seems on superficial examination to be complete; but even in the most affected limbs weak voluntary contractions may be detected, although they may not cause movement of the limbs.

If the disorder is not intense the hypotonia is the striking sign, while the weakness is noticed only by lack of energy in the movements. Only in one of Oppenheim's cases were the muscles of the trunk and neck affected, and the child, eight months old, could not sit up or hold the head upright. If the child were placed in a sitting posture the head, if not supported, fell backward.

Even when the myatonia is general the muscles of the eye, tongue, and throat escape, the diaphragm is normal, while the intercostal muscles seem to be involved.

The muscles feel flaccid, soft, and thin, although the term atrophy is not always suitable.

The electrical investigation, in the few cases in which it has been made, showed quantitative alteration, even to complete disappearance of the reaction. Only in one case could Oppenheim observe somewhat tardy contraction. In the mild cases the electrical reaction was normal.

Intelligence, sensation, and the special senses, so far as tests could be made at so early an age, showed no disturbance.

The condition appears to be always congenital, although it is not always noticed by the relatives of the child immediately after birth, and is just the opposite of Little's disease.

Oppenheim believed the pathology is to be sought in the muscles, and that these are arrested in their development. It is not probable, he thought in 1900, that the central nervous system is diseased.

He has seen improvement in some cases, and has been told of it by the relatives in others. He has never observed the condition in older children, and thinks it has no relation to progressive muscular dystrophy.

Disease of the cells of the anterior horns he does not exclude entirely. The affection may be confused with acute anterior poliomyelitis, but should be entirely separated from this disease. Anterior poliomyelitis develops acutely in a previously healthy child.

In his text-book he has referred to the disorder briefly.

Recently he has spoken again on this subject, In the report of the meeting of the Berliner Medicinische Gesellschaft, February 24, 1904, published in the *Berliner klinische Wochenschrift,* No. 10, March 7, 1904, p. 255, he says that the condition has excited little interest and that no necropsy in a case has been obtained. He has had four or five marked cases and a few abortive forms. Muscles innervated by cranial nerves always are intact and cerebral function is normal. The muscles are not really atrophic. He is unable to say whether complete recovery is possible. Hereditary tendency to the disease does not seem to have been observed in any of his cases. Because it is a congenital affection one might think of intrapartum hemorrhage of the spinal cord, but trauma at birth has not occurred in any case. Even as late as February, 1904, Oppenheim could find no mention in literature of the condition by anyone else. He refers to Vierordt's paper on pseudoparalysis of childhood depending on rachitis, syphilis, or Barlow's disease, in which hypotonia is a prominent symptom, but any of these conditions in being acquired is different from that described by Oppenheim.

Vierordt seems to explain the paralysis in his cases as a result of pain.

It is impossible to say from Oppenheim's papers whether there was a careful ophthalmological examination in all his cases or not. Sensation seems to have been normal, and pain does not occur, and the action of the sphincters is normal. As yet the subject has received only a hasty treatment from Oppenheim, but he has promised to discuss it more in detail.

A case of congenital myatonia with necropsy which I have had the opportunity to study, and which seems to belong to the type described by Oppenheim, is as follows:

B. T. was admitted to the University Hospital July 15, 1904, having been seen by me first in May, 1904. The patient was referred to me by Dr. A. Marcy, Jr., of Riverton, N. J. The history as taken on admission is as follows : The baby is twenty-two months of age.

Family History. The father and mother are living and well. One brother and three sisters are in good health. The mother has never had any miscarriages. The patient was born six years after the other children. There is possibly one case of epilepsy on the paternal side. No occurrence is known otherwise of heart, kidney, lung, nervous or neoplastic disease in the family, The father of this patient is said to be a "nervous" man. The mother is rather frail looking. There seems to be, therefore, no hereditary or family tendency toward the disease, and in this the malady is very different from progressive muscular dystrophy.

Previous Medical History. The baby was born at full term, and the birth was normal. It was breast-fed until the date of admission to the hospital. Nothing abnormal in the child was observed by his parents until he was about five months of age, when he did not seem to take notice of things as he should. Since this condition was noted he has apparently shown no change relative to sight. The anterior fontanelle closed at about the fifth month. Four months before admission the baby weighed twenty pounds. Since he was three or four months old he has been so constipated as to always require enemata. He cut his first tooth when about one year old, and now has ten teeth. When he was given cows' milk in an attempt to wean him he would always spit it out, and of late, since he has been taken from the breast, has swallowed with some difficulty. He has never at any time been able to hold anything in his hand. The mother has not noticed any increase in the child's weakness since his birth, and in this the disease is very different from amaurotic family idiocy.

An examination of the eyes made in Dr. de Schweinitz's service, May 7, 1904, is as follows: "Media clear, disk atrophic or (?) in right eye. The left disk could not be examined. No macular degeneration and no disseminated choroiditis. Some absorption of pigment. Convergent squint." Dr. Shumway thought the condition was not at all like that of amaurotic family idiocy.

Desiring to have further information regarding the eyes, I obtained the following letter from Dr. de Schweinitz:

"NOVEMBER 1, 1904.

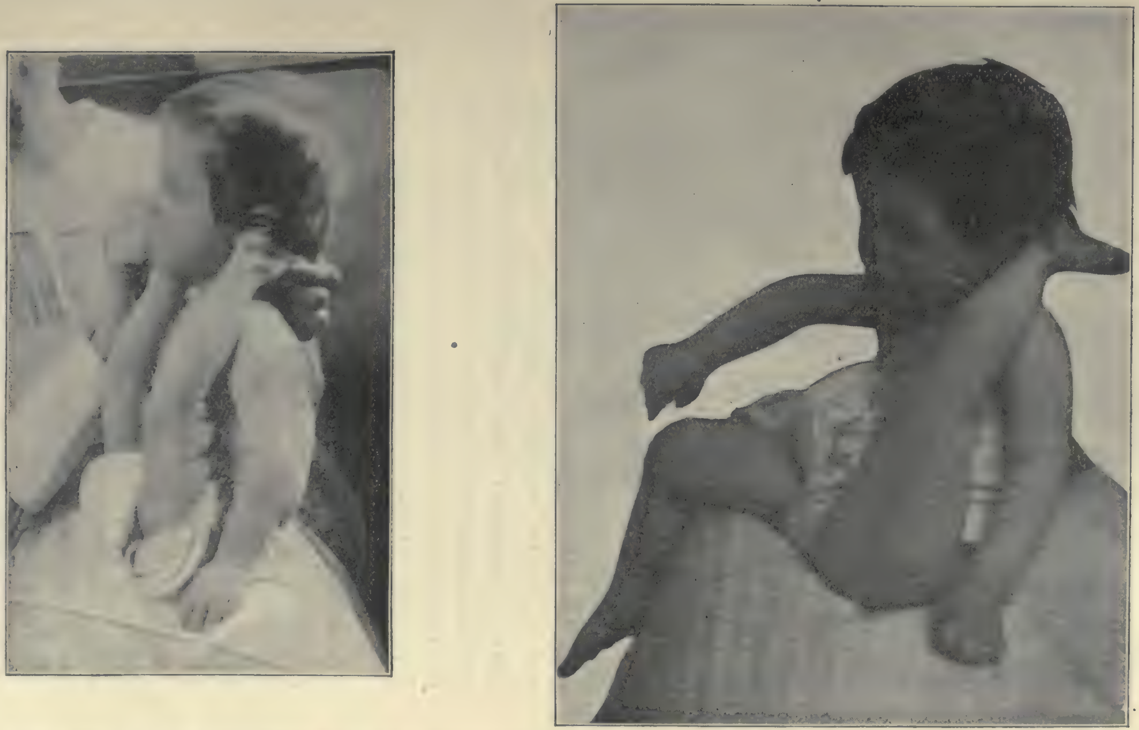
"MY DEAR DR. SPILLER: I regret to say that the case-books of the Eye Dispensary of the University Hospital do not contain any information in regard to the case concerning which you inquire. The name of the patient, without description of the ocular conditions, is all that is recorded. I presume that whoever made the examination forgot to transcribe the findings on the book. While I do not know what the missing word is, my impression is that it should be 'pallid'; in other words, that the sentence should read: 'Media clear, disk atrophic or pallid in the right eye.' This record is sometimes made when there is an uncertainty whether the pallor of the disk is pathological or not, pending future investigation or examination. Unfortunately, apparently neither the one nor the other was made, or if made was not recorded. I have a vague recollection of this child, and am inclined to think the examination was negative from the ocular standpoint; in other words, that there was no sign of organic change in the head of the optic nerve, but I am not sure of this. The words 'convergent squint' mean that the child had an ordinary concomitant convergent squint, and not an internal strabismus due to paralysis of the external rectus. Had it been the last-named condition the sentence would have been differently worded—that is to say, there would have been a definite statement that the squint was due to paralysis. I am sorry that I cannot give you more definite information.

Yours very truly,

"G. E. de Schweinitz."

I may add that my microscopic examination showed that the optic nerves were normal, and that at the necropsy there was no deviation of either eyeball.

Notes dictated by me May 10, 1904, are as follows: The child is fairly well developed for a baby of twenty months. The muscles do not appear to be wasted, and the upper and lower limbs are of good size, but the flesh is soft and flabby. The limbs are moved voluntarily and at all joints, and there is merely weakness present. The patellar reflex and Achilles tendon reflex are not obtained on either side, and any attempt to obtain the patellar reflex causes the child to draw up the lower limb on either side with considerable power, although not normally. Plantar irritation causes extension on the left side, but the movement is uncertain on the right side. Sticking any of the limbs with a pin causes the irritated limb to be drawn away with considerable force. The constipation is probably because of the atony of the abdominal walls. The leg can be hyperextended passively on each side, as shown in the accompanying illustrations. The



Figs. 1 and 2.—The photographs show the hypotonicity of the lower limbs. The front of the thigh could be placed in contact with the trunk, and the foot could be placed behind the head, without causing discomfort to the child.

hypotonicity of all the limbs, especially of the lower limbs, is very great. Either lower limb can easily be so flexed that the front of the thigh and leg is placed in close contact with the trunk, and the foot is placed behind the head without causing any discomfort to the child. The upper limbs are moved voluntarily, more freely than the lower. The child can sit alone only for a minute or two, and only when he is balanced, but his head falls slightly forward. He seems at times to notice a watch held before the eyes. He is, and always has been, unable to stand. The muscles of the limbs react to the faradic current. (Figs. 1 and 2.)

After the child was admitted to the hospital it gained one pound. It could not be induced to take food, and was always fed by the nasal tube. Its bowels moved voluntarily and it seemed to be improving. Its temperature was normal until August 1st, when it rose to 100.2°, but fell to normal the following day. On August 3d it rose to 102.4°, but fell to 99° on August 4th. On August 5th the temperature rose to 104° and the child died.

The necropsy was made by Dr. Coca August 6th, the following day. His report is as follows:

Clinical Diagnosis. Congenital hypotonia.

Anatomical Diagnosis. Fatty deposit in liver; tuberculosis of mesenteric lymph glands (?), Meckel's diverticulum.

General. Body of a white male child, weighing fourteen pounds; head relatively large; eyes sunken; there is no deviation of either eyeball; pupils unequal, left, 2.5 mm., right, 3.5 mm. in diameter. No rigor mortis is present; no post-mortem lividity. Joints are extremely mobile. Lower portion of abdomen shows post-mortem decomposition. *Preliminary incision*: Muscles light red, dry; extremely flabby. Fat of the calf is 6 mm. in thickness.

Abdomen. Peritoneum is smooth; large bowel is distended with gas. There are no adhesions. Very little fat is found in the omentum, which is short and free. Appendix measures 11 cm. At a point in the small intestine, 70 cm. from the ileocæcal valve, is found a diverticulum measuring 4 cm. in length.

Mesenteric lymph glands are much enlarged and whitish, some being pink in color. Some of these are preserved in Orth, others in 95 per cent. alcohol.

Liver weighs 240 grams; surface is smooth, mottled red and yellow; several irregular areas of pure yellow color are found, these extending for 5 or 6 mm. beneath the capsule into the organ. (Section in Orth.)

Gall-bladder contains a small amount of light, greenish-yellow bile.

Spleen weighs 12 or 15 grams; is dark red in color; free from adhesions; surface is smooth; consistency normal; Malpighian bodies can be readily seen; there are no gross lesions.

Pancreas 7 cm. in length; is soft; of a light pink color; shows no gross lesions. (Portion of tail taken for section.)

Kidneys. Capsule strips readily; stellate veins injected; cortex measures 4 cm. in thickness; no gross lesions.

Gastrointestinal Tract. Mucous membrane is thin; there is no congestion. Stomach contains considerable quantity of soft, greenish, mushy material.

Bladder is normal.

Thorax. Lungs. Both lungs are of a pale-pink color anteriorly; darker from congestion posteriorly. They are voluminous and crepitant everywhere. There are no adhesions.

Heart weighs 22 grams; pericardium is normal. Heart's flesh is pale and firm. Endocardium of both sides is perfectly normal. Foramen ovale is closed. Left ventricular wall measures 7 mm.

Thymus is present; shows no gross abnormalities.

Head. Brain and Cord. Taken for Dr. Spiller, together with muscle from the calf and arm, and nerves from the calf and the plantar surface of the foot.

Dr. T. H. Weisenburg was present at the necropsy and has furnished me with additional notes.

The hypotonicity of the limbs twenty hours after death was as great as during the life of the child, and the absence of post-mortem rigidity was striking. The muscles were flabby. In cutting into the muscles of the right calf the fat was found to be 6 mm. in thickness, there being very little muscle, and the muscle appeared paler than normal. There was hardly any muscle in the sole of the left foot where an incision was made in this part, and only fat with a little muscle could be removed for examination. A piece of the triceps muscle was removed on the left side. The subcutaneous fat over this muscle was 3 mm. in thickness, and the muscle was paler than normal.

The dura was adherent to the calvarium, so that the brain was removed with the calvarium attached. The brain weighed 980 grams.

The spinal cord and brain I found to be well developed, and the anterior and posterior spinal roots normal. Microscopic study of the brain, spinal cord, nerves, and muscles was made by me.

The nerve cells of the anterior horns of the cervical and lumbar regions are normal by the thionin stain. The anterior and posterior roots of the cervical and lumbar regions are normal. There is no spinal meningitis. The pyramidal tracts in the cervical and lumbar regions are not degenerated. Marchi sections of the cervical and lumbar regions show no degeneration.

The right and left optic nerves by the Weigert hæmatoxylin stain are normal, as are also Marchi sections of the left optic nerve.

Nerve cells of the paracentral lobules seem to be normal by the thionin stain, but Betz's cells are not numerous in the sections obtained. This does not indicate that they have degenerated.

A nerve from the upper part of the right upper limb stained by the Weigert hæmatoxylin or acid fuchsin with hæmalum is normal. The internal popliteal nerve and a nerve from the upper part of the right upper limb teased and stained by a 1 per cent. solution of osmic acid are normal.

Muscles from the sole of the left foot, from the back of the trunk, from the left calf, have a hyaloid appearance, and those from the sole of the foot are striking on account of the large amount of fatty connective tissue and on account of the considerable increase in the nuclei of the connective tissue. The muscle fibres are small, and those from the sole of the foot and from the left calf are much smaller than are those from the back of the trunk, and it is to be remembered that the child had more power in the muscles of its back than in those of the lower limbs. The transverse striations are well preserved, the longitudinal striations are not so distinct. Nerve fibres within the muscle from the sole of the foot appear to be normal.

This case may be regarded as of the type of congenital myatonia (not myotonia) described by Oppenheim. It is probable that when further cases are reported some modification of Oppenheim's description, perhaps only slight, may have to be made. He himself speaks of having observed some abortive forms of the disorder.

The parents of my patient never noticed any increase in the weakness of the child, and from the history there is reason to believe that the myatonia was congenital.

The microscopic examination shows that there was an arrest in the development of the muscle fibres, and that the central nervous system and peripheral nerves were normal. These findings explain the improvement that has been observed in some cases.

The congenital myatonia differs from progressive muscular dystrophy especially in the absence of family tendency, in being congenital, which muscular dystrophy commonly is not, and in the failure of progressive increase in the symptoms.

From anterior poliomyelitis it differs especially in that the latter begins acutely in a previously healthy child.

In amaurotic family idiocy the symptoms increase, the ophthalmological findings are pathognomonic, the disease occurs in more than one member of a family, and the pathology is very different.

Especially interesting in this case of congenital myatonia that I have been able to study were the persistence of the hypotonia (even twenty hours after death, as shown by the absence of post-mortem rigidity), the considerable amount of subcutaneous fat in the limbs, and the pallor of the muscles.

Congenital myatonia seems from the study of this case to be a muscular disease, and we may hope for improvement, at least in many cases, and this Oppenheim already has observed. As regards improvement the disease differs very greatly from amaurotic family idiocy, which is necessarily fatal. I[1] have recently had an opportunity to study the pathology of a case of amaurotic family idiocy, and the findings in that case and in this one of congenital myatonia are very different. I have made much effort to obtain muscle for comparison from a normal child of the same age as my patient, but have not succeeded.

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[1] American Journal of the Medical Sciences, January, 1905.

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CONGENITAL SPASTIC RIGIDITY OF THE LIMBS (CONGENITAL HYPERTONIA, LITTLE'S DISEASE).

REPORT OF TWO NEW CASES, WITH NECROPSY.

BY WILLIAM G. SPILLER, M.D.,
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(From the Neuropathological Laboratory of the University of
Pennsylvania and from the Philadelphia
General Hospital.)

IN discussing congenital spastic rigidity of the limbs, called especially by French writers Little's disease, it is well to have clearly in mind what Little[1] described in 1861, and therefore I quote lines taken from his original paper:

"Nearly twenty years ago, in a course of lectures published in the *Lancet*, and more fully in a *Treatise on Deformities*, published in 1853, I showed that premature birth, difficult labors, mechanical injuries during parturition to head and neck, where life had been saved, convulsions following the act of birth were apt to be succeeded by a determinate affection of the limbs of the child, which I designated spastic rigidity of the limbs of newborn children, spastic rigidity from asphyxia neonatorum, and assimilated it to the trismus nascentium and the universal spastic rigidity sometimes produced at later periods of existence.

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[1] Transactions of the Obstetrical Society of London, vol. iii. (for year 1861), 1862.

" . . . . It is obvious that the great majority of apparently stillborn infants, whose lives are saved by the attendant accoucheur, recover unharmed from that condition. I have, however, witnessed so many cases of deformity, mental and physical, traceable to causes operative at birth, that I consider the subject worthy the notice of the Obstetrical Society. In orthopædic practice alone, during about twenty years, I have met with probably two hundred cases of spastic rigidity from this cause. I omit reckoning the subjects of idiot and other asylums, in which probably such cases abound. . . . Occasionally the weakness of the limbs has been recognized as a genuine paralysis in the first instance, of which the rigidity of muscles has been the sequel. . . . Where the symptom of convulsions or 'inward convulsions' exists the rigidity is attributed to the convulsions. In many cases convulsions have been absent. . . . The greater or smaller impairment of intellect may safely be attributed to the greater or less mischief inflicted upon the cerebrum."

If there is still any doubt as to what Little had in mind the title of his paper removes it: "On the Influence of Abnormal Parturition, Difficult Labors, Premature Birth, and Asphyxia Neonatorum, on the Mental and Physical Condition of the Child, Especially in Relation to Deformities."

The definition given of Little's disease by Brissaud[1] is "spastic and congenital paraplegia of the four limbs, more pronounced in the inferior, occurring especially in children born before full term, characterized by spasm more than by paralysis, and not associated with convulsions or disturbance of intellect, and capable of progressive amelioration, if not of complete cure."

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[1] Maladies Nerveuses, p. 110.

This is a very different description from that given by Little himself, who recognized feeble mentality and convulsions as part of his symptom-complex.

Van Gehuchten[1] makes another distinction in that he would limit the term Little's disease to cases of premature birth with spasticity. The importance of a separation of the cases resulting from premature birth from cases of spasticity developing after an injury to the brain should be recogsized, because there is more hope for improvement where the motor tracts have been arrested in their downward growth into the cord by premature birth, but there is no good reason why these cases alone should be entitled Little's disease.

It is not my intention at this time to discuss fully this symptom-complex, as I have already written upon the subject, but rather to refer to the pathology of these cases of congenital spasticity of the limbs. Freud, Brissaud, Marie, Van Gehuchten, B. Sachs, Dejerine, and others have presented the symptomatology clearly, and there is little left to say. An interesting discussion on the subject has been held recently by the Neurological Society of Paris.[2] Some would exclude the spinal form of congenital spastic rigidity, not because of the symptom-complex, but because the lesions are spinal. At present, cases of congenital spastic rigidity without paralysis, cases with paralysis, cases with epilepsy, cases with feebleness of intellect, cases resulting from injury of the brain at birth, cases resulting from premature birth and thereby arrest in the development of the central motor fibres, cases of cerebral lesions, and cases of spinal lesion are designated Little's disease, and so great is the confusion that it has been proposed to have a congress of physicians determine what shall be included under this designation. As in progressive muscular atrophy of the Aran-Duchenne type different affections were included, and we now honor Duchenne by recognizing only one disease as representing this type, so it is hoped Little's name may be preserved by excluding some of the different forms of congenital spastic rigidity now classed under the term Little's disease. At the same time it should be remembered that Little confused many types.

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[1] Journal de Neurologie, 1896.

[2] Revue Neurologique, June 30, 1903, p. 640.

The cases of congenital spasticity without macroscopic lesions are not very numerous.

Ganghofner[1] reported a case of what was apparently general spasticity. Sections from the upper part of one anterior central gyrus appeared normal, but a scarcity of fibres in the crossed pyramidal tracts of the cord was noticed. Macroscopically the brain and cord were normal, and yet he believed that the cortex could not really have been perfectly developed. In two other cases of spasticity in which no peculiar findings were observed by the microscope, he thought there might have been a moderate decrease of the pyramidal fibres which eluded detection. I referred to these cases in my former paper on Little's disease.

Rolly reported two cases of congenital spastic rigidity in twins. The father had syphilis. One child was asphyxiated and convulsions began on the eleventh day, and lasted about four weeks with intermissions. The other child had convulsions also. Both children died in the first month of life. In both cases spasticity existed without paralysis, and in both athetosis was seen. In both cases the neuroglia and bloodvessels were proliferated, and in one case the nerve cells of the spinal cord and brain were much altered. Macroscopically little of importance was seen. The pyramidal tracts were normal in each case, therefore Rolly thinks these cases support the view that the cause of congenital spasticity is to be found in alteration of the cells, and not in alteration of the pyramidal tracts, and yet the method of Marchi does not appear to have been employed. It might be difficult to detect a slight degeneration of the pyramidal tracts by Weigert's hæmatoxylin method within the first months of life, as at this period these tracts are imperfectly medullated, and the alteration by the Marchi method would depend on the age of the child at death.

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[1] Zeitschrift für Heilkunde, 1896, vol. xvii.

In another paper he reported two more cases of congenital spastic rigidity without paralysis. Macroscopically little that was abnormal was to be seen in these cases also. In both cases there was proliferation of neuroglia and of bloodvessels and leptomeningitis.

Rolly[1] finds it very difficult to separate the cases of congenital spasticity without paralysis in the literature from those with paralysis, because the types are not sharply drawn. It is doubtful whether this distinction can be maintained, as the weakness may in some cases be very slight and in others more pronounced.

Dejerine a few years ago remarked in his *Sémiologie du système nerveux*, p. 552, that no necropsy in a case of Little's disease had been published demonstrating a primary agenesia of the pyramidal tracts. In most of the reported cases hydrocephalus, porencephaly, or cerebral sclerosis had been found as the primary lesion.

In February, 1898, I[2] reported two cases of congenital spastic rigidity, with necropsy.

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[1] Deutsche Zeitschrift für Nervenheilkunde, October 24, 1901, vol. xx., Nos. 3 and 4.

[2] Journal of Nervous and Mental Disease 1898, p. 81.

Case I. was that of a girl, Kate K., born in difficult labor at full term. The head was small, fourteen and a quarter inches in circumference when the child was nineteen months old. At this period she was unable to sit without support, but could hold objects in her hands. She had moderate contracture of the flexor muscles of the feet, but the lower limbs were not paralyzed. The child was an idiot. She had convulsions when teething. She died when six years old. When the child was examined after death by me the feet were in the position of marked talipes equino-varus and the toes were flexed. The circumference of the head was sixteen inches. The brain was small and weighed sixteen ounces. There were no abnormal depressions, no sclerotic areas, and the gyri and fissures were well developed; in brief, there were no gross lesions. The cord was small. The cells of Betz in the paracentral lobules were not numerous, and those present did not seem to be so large as those in normal brains. The nerve cells of the anterior horns of the spinal cord were not in the usual number, but especially remarkable was the fineness of the nerve fibres forming the crossed pyramidal tracts. There was evidently an arrest in the development of these fibres.

Case II.—R. C., eight years old, was an idiot. He had never spoken, and did not understand what was said to him, and took no notice of anything. The limbs and trunk were well nourished. He could not walk and made no attempt to take a step. In the erect position he inclined backward, and had to be supported or would fall. He could move his legs when sitting. The patellar reflexes and Achilles tendon reflexes were exaggerated, and slight ankle clonus was present. The movements of the upper limbs were good, and the reflexes of these limbs were not distinctly exaggerated. Strabismus was not observed, and the pupils were equal. The head was well developed. This child was born at full term and in easy labor. A few months after birth he had convulsions. His condition was congenital. My recollection is that this boy was spastic; although spasticity is not distinctly mentioned in the notes, it is indicated by his inability to move his lower limbs while standing and by his tending to fall backward while standing.

Three or four months after I made the clinical examination just referred to, the boy died in status epilepticus. The brain presented no gross lesions. I was unable to find any very positive microscopic lesions. It was impossible to detect any diminution of the fibres in the crossed pyramidal tracts, and the fibres in these tracts were not very fine. The nerve cells of the anterior horns of the cervical and lumbar regions were numerous and well formed. The only explanation I can offer for this case is that the nerve fibres in the pyramidal tracts had not grown downward into the cord in normal number so far as the lumbar region, although the cervical region was well supplied. It would be very difficult to establish the correctness of this view.

In 1897 Dejerine showed that congenital spastic rigidity might be caused by spinal lesions. In his case a sclerotic focus was found in each crossed pyramidal tract at the second cervical segment. Recently he has reported a similar case. A man, aged sixty-six years, born at full term, had congenital spastic rigidity of all four limbs. Contracture was very marked, especially in the lower limbs. Intelligence was good. The brain was normal. A sclerotic patch was found in the spinal cord in the third cervical segment, and this had caused secondary degeneration.

Both of these cases were regarded by Dejerine, before necropsies were obtained, as being of cerebral origin. The spasticity was supposed to be the result of a lesion in each cerebral hemisphere. The cases show that congenital spastic rigidity may be of spinal origin, and probably also that myelitis may occur in the child in utero. The latter is a very important conclusion.

Is it possible to distinguish clinically between the cerebral and spinal forms of congenital spastic rigidity? Intelligence below the average, paresis and spasm in the facial muscles, and convulsions with increased tonicity of the limbs are indicative of the cerebral form, but these signs may be absent and the lesion still be cerebral if the upper limbs are not spastic. The absence of these signs in a case of great spasticity of all the limbs, according to Dejerine,[1] implies that the lesion is in the cervical region of the spinal cord, presumably because a cerebral lesion sufficiently intense to cause spasticity of all the limbs would be likely to cause feeble intellect, paresis and spasms of the facial muscles, and convulsions of the limbs.

I have had, since the publication of my former paper, two cases of congenital spastic rigidity with necropsy, and the second one (Case IV.) is especially interesting in connection with the two cases of Dejerine referred to above. It shows how difficult it may be to distinguish the cerebral from the spinal form during the life of the patient. In this case there was feeble mentality with spasticity of all the limbs, but the cause of the spasticity was found in a compression of the cervical portion of the cord by displaced vertebræ.

Case III. shows the arrest in the development of the pyramidal fibres like that previously reported as occurring in Case I.

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[1] Revue Neurologique, June 30, 1903.

CASE III.—The child was brought to me by its parents about 1899. I made a careful examination of the patient at that time, and gave my opinion that the case was a typical one of congenital spasticity of the limbs. Unfortunately I kept no notes of the examination. The following report has been obtained from the father of the child, who is a man of unusual intelligence, and capable from his training and position of appreciating the importance of accuracy in the report.

R., first child in the family, female, was born November 1, 1897, and died May 7, 1900, at the age of two years and six months. The mother was twenty-nine years old at the time of the child's birth; the father was twenty-nine and a half years old.

The heredity of the mother was good. The father had nine paternal aunts and uncles, and one aunt and one uncle had mental disease. The uncle had two attacks of suicidal melancholia, the aunt had paranoia.

R. was born with the help of forceps after a pregnancy of seven months. During the pregnancy the mother had hemorrhage twice, presumably from the uterus.

The child weighed at birth three and a half pounds, and when eight months old weighed eleven and a half pounds. She was nourished at the breast during the first two months.

Jaundice developed in the first week after birth, and lasted three weeks. She had bronchitis three times, and frequently had disorders of digestion. She never slept well, and sleeplessness was more marked as she grew older. During the last months of life she awoke about every two hours.

When she was a year and two or three months old she had a convulsion. The epileptic convulsions became more and more frequent, until during the last months of life she had at times as many as three a day. The whole body was implicated in the spasmodic movements and consciousness was lost.

The child was never able to walk. She would hold for a little while things put into her hands, but she never learned voluntarily to bring her hands in contact with objects she wanted. She, however, made inadequate movements. She could not sit up nor hold her head up, and she kept her mouth open much of the time.

She never learned to speak, not even to say "papa" or "mamma." She made some of the sounds which usually precede the first words.

She had associated going out of the house with being dressed, or rather with almost any change in her clothing. She had also associated the wearing of a hat by her mother or by her nurse with going out. On these occasions she expressed joy. She never learned to distinguish between the clothing put on to go out and other changes made in her dress.

Joy and pain were the only emotions she manifested. An instinctive fear-reaction, quite meaningless to her, was observed when she was nine months old. It was caused by a shrill noise made by an iron hook on falling. She had no idea whatever of danger at the time, neither had she at any later time, so far as could be determined.

Two children, a boy and a girl, born subsequently to R., are well developed mentally and physically.

A necropsy was made in the case by Dr. T. F. Branson, in whose care she had been, and myself. The brain appeared to be well developed. Microscopic examination of the brain and cord by me showed numerous Betz cells in the paracentral lobule. They stained diffusely with thionin, and the chromophilic elements were not distinct. This may have been a post-mortem change. The nerve cells of the anterior horns of the cervical and lumbar regions were numerous and appeared to be normal. The nerve fibres in the crossed pyramidal tracts of the cervical region were very fine, and finer than those of the columns of Goll, and finer than the fibres of the crossed pyramidal tracts from a normal child of the same age. The Weigert hæmatoxylin stain showed no degeneration.

It is desirable to emphasize the findings in this case. There was evidently an agenesia of the pyramidal tracts in the spinal cord without macroscopic lesions, as in my first case, and the cause was probably premature birth at the seventh month. The feeble mentality and convulsions may have been caused by the difficult labor. Convulsions are not usually caused by premature birth. The brain of this child was well developed.

Case IV.—Euphemia F., aged seventy years, was admitted to the Philadelphia General Hospital, December 18, 1897. On admission she had loss of power in the upper and lower limbs and difficulty in speech. Her family history was negative. It was impossible to obtain much history from the patient. She had had scarlet fever and measles in childhood. She had always been unable to use the lower limbs at all, but had some power of movement in the upper limbs. She said she had been born in this condition.

Notes dictated by me, June 3, 1903, are as follows: She is unable to talk. When asked her name she mumbles and it is impossible to understand her. The tongue is protruded straight, it is not atrophied, and is without fibrillary tremors. The facial muscles do not appear to be involved. The masseter contracts well on each side. She moves the head freely from side to side. She understands what is said to her, but her mentality is feeble.

The whole body is poorly developed. The movements of the upper limbs are awkward and very limited. Each forearm is contractured on the arm in an obtuse angle. The left wrist is not contractured. There is a slight tendency to flexor contracture of the left fingers, but this is readily overcome. The right hand is contractured in flexion at the wrist. The fingers of the right hand are contractured in flexion, especially the ring finger. The thumb is straight. It is impossible to test sensation accurately, but when the upper limbs are touched with a pin she makes a slight effort to withdraw them. The biceps tendon, triceps tendon, and wrist reflexes are not obtained distinctly on either side because of muscular atrophy and contracture. She has, however, a slight biceps response on the left side.

The lower limbs are very rigid. The thighs are partly contractured on the abdomen and the legs on the thighs. The feet are œdematous and the big toe on each side is hyperextended. The patellar reflex is exaggerated on each side. Ankle clonus is not obtained on either side. The Achilles tendon reflex is obtained on each side, on the left side there is some movement of the foot, but on the right side the reflex is shown only by contraction of the calf muscles. Babinski's reflex is very marked on each side. Voluntary movements of the lower limbs are possible but are feeble. Pin pricks are perceived in the lower limbs, as shown by withdrawal of the limbs when they are pricked.

The heart is weak but regular. There is a systolic murmur. The pulse is feeble.

The patient died June 5, 1903.

A post-mortem examination was made the same day. The brain was œdematous and the ventricles contained a somewhat greater amount of fluid than is usually found. The post-central gyrus on each side in its upper part was small. The bodies of three or four upper cervical vertebræ extended backward into the spinal canal and compressed the spinal cord partially. The cervical cord in the compressed portion was considerably smaller than in the thoracic portion, and the posterior columns appeared degenerated. A microscopic examination of the spinal cord was made.

Fourth cervical segment. (Fig. 1.) The direct cerebellar tract and Gowers' tract were much degenerated on the right side, but the degeneration extended farther inward than the area of Gowers' tract. The direct cerebellar tract on the left side was not degenerated, but Gowers' tract and the area internal to it were as much degenerated on the left side as on the right. The crossed and direct pyramidal tracts were not degenerated at all at this level. The posterior columns were greatly degenerated, but there was an area of normal fibres on the inner side

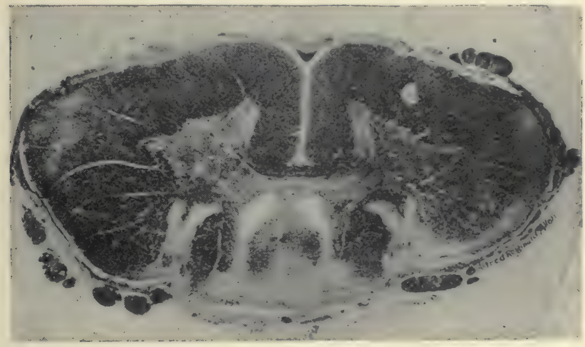

Fig. 1.—Section from the fourth cervical segment (Case 4), stained by the Weigert hæmatoxylin method. The direct cerebellar tract and Gowers' tract on the right side, as well as Gowers' tract on the left side, are degenerated. The posterior columns are much degenerated.

of each posterior horn, showing that normal posterior roots entered above the area of compression. Some normal fibres also were found in about the centre of each column of Goll.

Sixth cervical segment. (Fig. 2.) The posterior columns at this level were almost destroyed. Some normal fibres were found in the area of the right posterior roots at this level, but very few normal fibres were found near the left posterior horn. The right direct cerebellar tract and Gowers' tract were much degenerated, as were also the left Gowers' tract, but not the left direct cerebellar tract, unless in its ventral portion. The nerve cells of the anterior horns were smaller than normal, their processes were imperfect and many cells contained few chromophilic elements but contained much pale yellow, granular material. There was no meningitis.

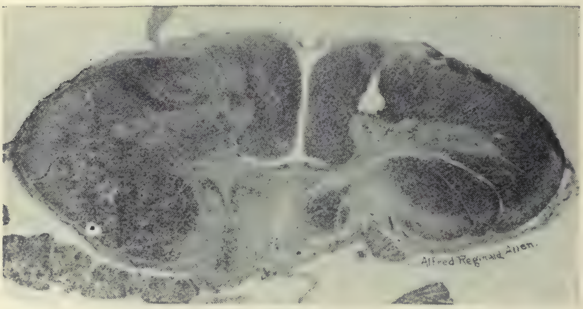

Fig. 2.—Section from the sixth cervical segment (Case 4) stained by the Weigert hæmatoxylin stain. The posterior columns are almost destroyed. The right direct cerebellar tract and Gowers' tract are degenerated, as is also the left Gowers' tract. The anterior columns are not normal.

Eighth cervical segment. (Fig. 3.) The degeneration in the posterior columns was confined to Schultze's comma zones. The right crossed pyramidal tract was degenerated in its posterior portion. The right direct cerebellar and right Gowers' tract were moderately degenerated, but not so much as at higher levels. The left Gowers' tract was slightly degenerated, but not the left direct cerebellar tract. The left crossed pyramidal tract did not present much degeneration.

Fourth thoracic segment. The degeneration of Schultze's comma zone on each side at this level was scarcely indicated. The posterior part of the right crossed pyramidal tract was degenerated. The degeneration of Gowers' tract on each side was slight, and the right direct cerebellar tract was scarcely degenerated.

The nerve cells of the anterior horns in the lumbar region were in a much better condition than those of the cervical region. They contained much pale-yellow, granular matter. Here and there a degenerated nerve cell was found.

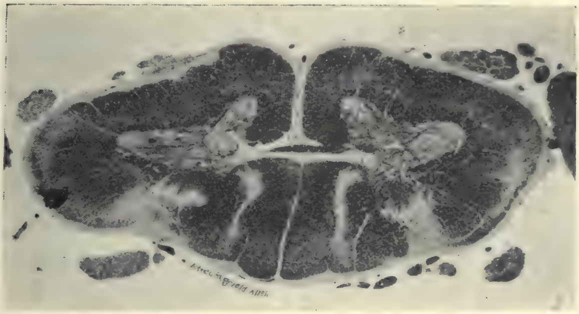

FIG. 3.—Eighth cervical to first thoracic segment (Case 4). The degeneration in the posterior columns is confined to Schultze's comma zones. The right crossed pyramidal tract is degenerated in its posterior portion. The right direct cerebellar and right and left Gowers' tracts are moderately degenerated.

Sections from the medulla oblongata showed no very striking changes.

Amyloid bodies were very numerous in the outer portion of the cortex of one of the paracentral lobules, which was not remarkable, as the woman was seventy years old. The Betz cells were well formed.

The degeneration of the right crossed pyramidal tract in its posterior portion, although comparatively slight, was sufficient to explain the spasticity and paralysis of the limbs of the right side, but the condition of the left crossed pyramidal tract was hardly adequate to explain these symptoms in so far as they related to the left limbs. The degree of degeneration could only be determined by the amount of neurogliar proliferation, as the Marchi method was not applicable, and where a lesion occurs early in life, especially if it is congenital, and the patient lives to adult age, the proliferation may be very slight, as I have seen in other cases.

The feeble mentality and disturbance of speech with the spasticity of all the limbs suggested a cerebral lesion as the cause of the symptoms; but there was sufficient in the spinal cord to explain the spasticity, and the case may be regarded as one of the rare examples of early spasticity of all four limbs caused by a lesion of the spinal cord in the cervical region. The small size of the upper part of each post-central convolution can hardly be regarded as explanatory of the spasticity if we accept the opinion of Grünbaum and Sherrington, who place the motor area entirely in front of the Rolandic fissure.

The cause of the displacement of the vertebræ could not be determined. It may have been caused by pulling on the head during childbirth, but there is no way in which this may be known. The condition of the posterior columns in the area of the vertebral displacement indicates that the lesion occurred early in life. The cord seems to have been arrested in its growth chiefly in the posterior columns, and the small size of the posterior columns, although they are almost entirely sclerotic, shows that they were damaged before they had become well developed.

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PSEUDOBULBAR PALSY.[1]

REPORT OF THREE CASES WITH NECROPSY AND OF THREE CASES WITHOUT NECROPSY.

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(From the Neuropathological Laboratory of the University
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ALTHOUGH the symptom-complex of pseudobulbar palsy has been well recognized, especially in the German and French literature, it is remarkable that, with the exception of one clinical case reported within the past year by Hirschberg in this country, and of a case of infantile pseudobulbar paralysis reported by Marshall and Jacob in England, there is no mention of this symptom-complex in the English literature, so far as I know. It is probably not because it is infrequent, for Urstein, in his monograph on pseudobulbar palsy, published in 1900, was able to collect one hundred and fifty cases, eighty-six of these being with autopsy, but because it has not been sufficiently differentiated from the other forms of bulbar paralysis. Therefore, the following report of six cases, three with microscopic examination, should be of interest:

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[1] Read before the Philadelphia Neurological Society, December 27, 1904.

Case I.—B. P., aged fifty-six years, teamster, admitted to the wards of the Philadelphia General Hospital, June 14, 1900. The family history is unimportant. The patient had the ordinary diseases of childhood, rheumatism, smallpox, gonorrhœa, and three or four chancres. He was not an alcoholic.

One year before his admission to the hospital he had middle-ear disease, which resulted in a left-sided superficial facial palsy and deafness on that side. A few months after that, while walking, he suddenly felt dizzy and weak, and had to be carried home. Examination at that time showed a left-sided paresis of the arm and leg. He also said that at that time his speech became slow and his voice was not as distinct as formerly. His memory also became poor. A short time after the first attack of hemiplegia he had a second attack, which again involved the left side of the body, causing a paresis of the left arm and leg.

Examination at the time of his admission to the hospital, which was about five months after the second hemiplegic attack, showed a paresis of the left arm and leg, and a complete superficial paralysis of the left side of the face. The tongue deviated to the left, the pupils were unequal, and the tendon reflexes of both upper and lower limbs were much increased. The Babinski reflex was obtained only on the left side.

Eight months after the examination the patient suddenly became confused, slipped over to his right side while in his chair, and was put to bed in an unconscious condition. The head and eyes were strongly deviated to the right. He recovered in a few days, and it was found that his speech was more indistinct than before.

Examination by Dr. Spiller while the patient was in his service, made on July 27, 1901, a little over a year after his admission to the hospital, was as follows:

"The man presents the appearance of pseudobulbar palsy. In eating he chokes often. In feeding himself he places his mouth close to the dish, so as to carry the food as short a distance as possible. He chokes oftener when fed than when he feeds himself. He has frequent attacks of crying, and saliva constantly dribbles from his mouth. It is impossible to make him talk, for he is very deaf and demented, though it is said that he is able to talk with his relatives, but his speech is understood with great difficulty. In the attacks of involuntary crying the left side of the mouth is drawn higher than the right. He chokes often, even when not taking food. (Fig. 1.)

"He cannot stand at all. The left side of the face is smoother than the right, and the forehead wrinkles only on the right side. There is no ptosis. The tongue is protruded about a normal distance, and in the median line, and there is no fibrillary tremor or atrophy of the tongue.

"*Upper Limbs.* The right upper limb is spastic, but not intensely so. Voluntary movements are limited. The left upper limb is very spastic and is almost completely paralyzed. The biceps, triceps, and wrist reflexes are exaggerated on both sides, more so on the left.

"*Lower Limbs.* Both lower limbs are spastic, the left more so. The patellar reflexes are exaggerated on each side. The Achilles jerks are not obtained, probably on account of the spasticity. Ankle clonus is not present on either side. Sensation cannot be tested on account of the dementia, but when the patient is stuck with a pin he makes some feeble movement of defence. There is incontinence of urine and feces."

The man died soon afterward in a uræmic condition. The autopsy was performed by Dr. Simon Flexner, and the following conditions were found: "Acute splenic tumor, lobular and pseudo-pneumonia, emphysema of the lungs, moderate chronic nephritis, and arteriosclerosis of the cerebral vessels.

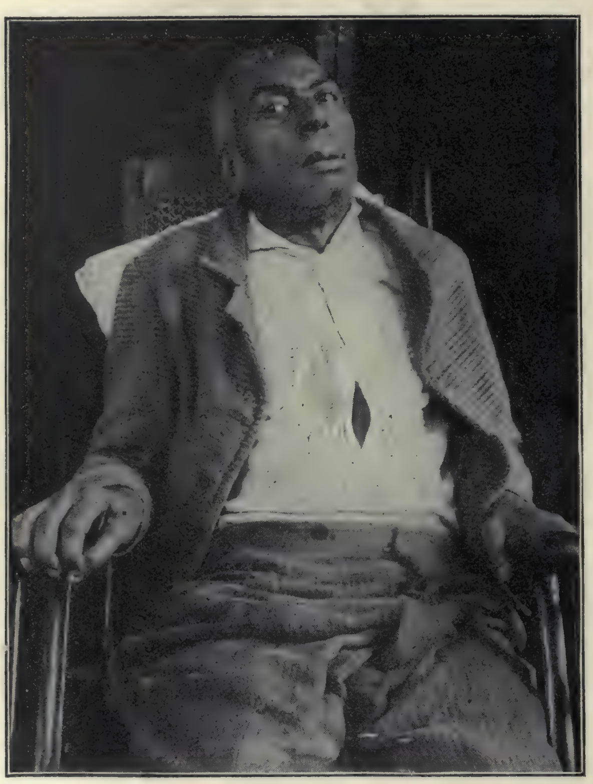

FIG. 3.—The photograph shows the man (Case I.) as he appeared constantly. There was never any change of expression.

"MACROSCOPIC EXAMINATION. An area of sclerosis about 20 mm. anteroposteriorly and about 15 mm. laterally is found in the lower portion of the left cerebellar lobe. Another and smaller area is found posterior to that just described, in the posterior part of the left cerebellar lobe. The bloodvessels at the base of the brain are very sclerotic. Two small tumor-like formations, each about the size of a split pea, are found in the under surface of the corpus callosum, directly back of the quadrigeminal body. These are of brownish-gray color.

"The ventricles are unusually distended with fluid. An area of softening is found in the anterior portion of the posterior limit of the left internal capsule. Small cysts are found in the choroid plexus."

MICROSCOPIC EXAMINATION. *Lumbar Region.* By the Weigert hæmatoxylin method there is a faint, almost imperceptible degeneration in the left crossed pyramidal tract. At this level there is a slight round-cell infiltration of the pia. The nerve cells of the anterior horns, as shown by the thionin stain, are not distinctly degenerated. The Marchi method shows no recent degeneration in the lumbar and sacral regions.

Midthoracic Region. The left crossed pyramidal tract shows a very slight, almost imperceptible degeneration, but the direct pyramidal tracts, especially the right, are distinctly degenerated. The right direct and the left crossed pyramidal tracts show slight recent degeneration by the Marchi method. There is a slight round-cell infiltration of the pia in certain parts.

Cervical Region. (Fig. 2.) Sections through the lower part of the cervical swelling show a very marked degeneration of each direct pyramidal tract, more distinct in the right. It extends from the anterior periphery of the cord to the white commissure. The left crossed pyramidal tract is slightly degenerated. Both direct pyramidal tracts, especially the right, and the left crossed pyramidal tract show moderate recent degeneration by the Marchi method. The nerve cells of the anterior horn are in a fair state of preservation, although occasionally a diseased cell is found. Round-cell infiltration, as in the lower levels of the spinal cord, is of moderate degree.

Medulla Oblongata. The right anterior pyramid is distinctly but not intensely degenerated. The most external portion of the left anterior pyramid is also slightly degenerated. The round-cell infiltration of the pia is more intense here than

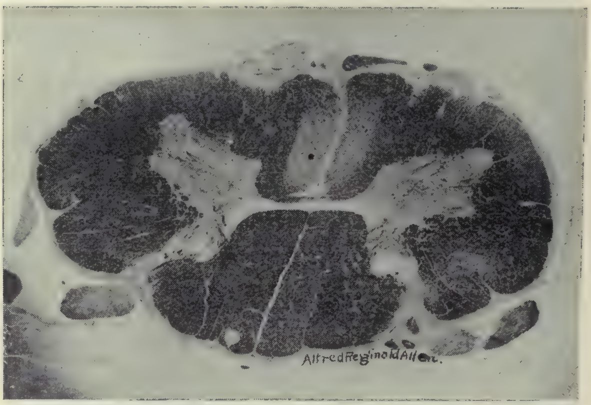

FIG. 2.—Section from the lower cervical region stained by the Weigert hæmatoxylin method (Case 1). Both direct pyramidal tracts are much degenerated, and the left crossed pyramidal tract (the photograph shows the section reversed) is slightly degenerated. The degeneration of the direct pyramidal tracts extends to the periphery of the cord, although in the photograph it does not appear to do so, because the section was a little thicker at the periphery.

in the spinal cord, and there is also a slight perivascular round-cell infiltration. The cells of the nuclei of the twelfth nerves seem to be normal. The intramedullary fibres of the hypoglossal nerves are normal. The cells of the nucleus ambiguus of each side are normal, as are also the cells of the nuclei of both facial nerves, as seen by the thionin stain.

Pons. The right pyramidal tract in the pons is partially degenerated. The left appears to be normal, except in one or two bundles quite near the raphe and in the ventral part of the pons.

Cerebral Peduncles. (Fig. 3.) A narrow band of degeneration is found near the middle of the foot of the right cerebral peduncle. The innermost portion of the foot of the left cerebral peduncle is partially degenerated. There is a slight round-cell infiltration of the pia around the cerebral peduncles.

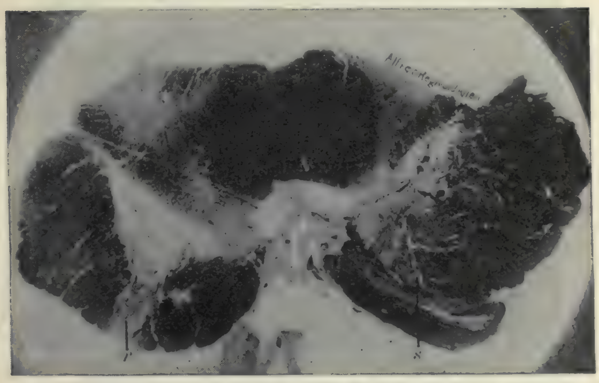

Fig. 3.—Section from the cerebral peduncles (Case 1), showing degeneration of the middle part (*r*) of the right foot and of the median bundle (*s*) of the left foot. The latter is not very clearly shown in the photograph.

In the right internal capsule, directly in the middle of the posterior limb, is an area of degeneration occupying approximately about one-fifth or one-sixth of the entire length of the posterior limb. An old area of softening is found in the external division of the lenticular nucleus, but this could not have caused the degeneration of the middle part of the posterior limb of the internal capsule. (Fig. 4.)

On the left side of the brain an old area of softening is found in the anterior portion of the optic thalamus, extending as high as the upper portion of the internal capsule. The fibres in the internal capsule in the most anterior portion of the posterior limb—*i. e.*, at the knee—are partially involved in this area. (Fig. 5.)

The sections show that a degeneration confined to the middle portion of the posterior limb of the internal capsule, as in the right side of this brain, causes a degeneration of the middle portion of the foot of the cerebral peduncle, and that a degeneration confined to the most anterior portion of the posterior limb of the internal capsule, as in the left side in this brain, causes a degeneration of the innermost portion of the foot of the cerebral peduncle.

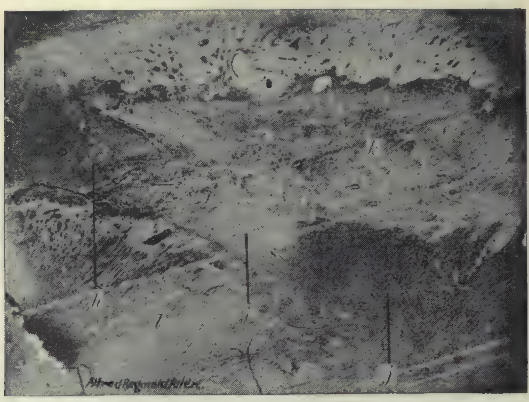

Fig. 4.—Section stained by the Weigert hæmatoxylin method (Case 1). *h*, posterior part of right posterior limb of the internal capsule; *i*, area of degeneration in the middle part of the posterior limb of the internal capsule; *j*, knee of the internal capsule; *k*, lenticular nucleus; *l*, optic thalamus. The upper portion of the photograph, not designated by a letter, represents an area of degeneration in the outer part of the lenticular nucleus.

Cranial Nerves. Sections of both optic nerves stained by the Weigert hæmatoxylin and acid fuchsin methods show nothing abnormal. A cross-section of the optic chiasm is normal. A moderate round-cell infiltration is seen in the pia about the chiasm. Transverse and longitudinal sections of either oculomotor nerve show nothing abnormal.

The tongue is not altered, and the nerve fibres within the tongue are normal.

The muscles of both vocal cords are perfectly normal, and the nerves within these muscles are also normal. Sections of these muscles stained by the Marchi method show no recent degeneration.

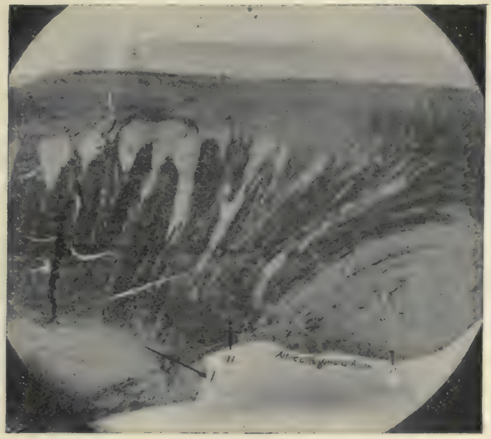

Fig. 5.—Section of left internal capsule, showing area of degeneration in knee (u) and anterior part of optic thalamus (l).

Summary. Man, aged fifty-six years, syphilitic. History of several attacks of hemiplegia involving the left side of the body, but no distinct history of a right hemiplegia, although the patient had subsequently slight attacks, and the examination showed a weakness of all the extremities, distinctly more marked, however, on the left side. The reflexes were also more exaggerated on the left side. Sensation was normal. Speech was bulbar in character; there was difficulty in swallowing and eating; dribbling of saliva, forced crying, and dementia.

Microscopically, on the right side an area of secondary degeneration was found in the middle part of the posterior limb of the internal capsule, and a sclerotic area in the outer portion of the lenticular nucleus. The middle part of the foot of the right cerebral peduncle was degenerated. The degeneration of the motor fibres was traced through the pons and medulla oblongata, and into the lumbar region of the spinal cord. In the cervical and thoracic regions the degeneration of the right direct pyramidal tract was more intense than that of the left crossed pyramidal tract. On the left side of the brain an area of softening was found in the knee of the internal capsule. The innermost part of the foot of the left cerebral peduncle was degenerated. The degeneration in the pons was limited to one or two bundles quite near the raphe and in the ventral part of the pons. In the medulla oblongata it was confined to the outer part of the pyramidal tract. In the cervical and in the midthoracic regions the degeneration was confined to the direct pyramidal tract. The nuclei and the intramedullary fibres of the different cranial nerves were normal. The optic and oculomotor nerves were normal. The tongue and the laryngeal muscles were normal. Syphilitic round-cell infiltration of the pia was apparent in the spinal cord and brain.

Case II.—W. W., aged fifty-two years, baker; admitted to the nervous wards of the Philadelphia General Hospital on July 18, 1904, in the service of Dr. Charles S. Potts. The family history is negative, and the patient's past history is unimportant. The patient's wife and son-in-law state that four years ago he had a left-sided hemiplegia.

This improved considerably. One year afterward he suddenly became unconscious, and had a paralysis of the right side of the body. After this second attack, the condition of his left arm and leg improved so that he could walk with the aid of a cane, but the right arm and leg remained paralyzed. He had no difficulty in speaking until after the second attack, when his speech became slow and indistinct, so that at times his attendants could not understand him; he also had difficulty in swallowing, sometimes choking even on drinking water. Saliva also constantly dribbled from his mouth. His wife also states that, although after the second attack the left arm and leg improved for a short time, they have since become weak. For the past six months he has not been able to control his urine. Memory is fair. The report of the examination by Dr. Potts on July 20, 1904, is as follows:

"The pupils are small and equal and respond promptly to light, accommodation, and convergence. The left side of the face droops and the mouth is drawn to the right, and the patient cannot whistle. The nasolabial fold is more pronounced on the right side. The tongue is protruded faintly to the left.

"*Upper Limbs.* There is slight power of flexion in the right arm, and the grasp is feeble, no other movements being possible. The left upper limb is almost completely paralyzed, and there is some slight rigidity of the left elbow. The biceps, triceps, and wrist reflexes are prompter than normal on both sides, especially on the left side. Sensation for touch, pain, and temperature is normal.

"*Lower Limbs.* Both lower limbs are weak, the left one probably a little more so. The patellar and Achilles jerks are prompter than normal, especially on the left. Ankle clonus is obtained on the right side, but not on the left. Babinski's phenomenon is obtained on both sides. Sensation for all forms is normal.

"There is no incontinence of urine and feces. The patient is stupid, and it is difficult at times for him to understand commands. The radial arteries are tortuous and cord-like. The urine examination is normal."

The patient died on August 1, 1904. The autopsy was performed by Dr. Guthrie McConnell. The following conditions were found:

Chronic obliterative pleuritis of the right side, hypertrophied heart, mitroendocarditis, atheroma and sclerosis of the aorta and coronary arteries, chronic parenchymatous nephritis, and congestion of the liver.

MACROSCOPIC EXAMINATION. An old sclerotic area directly at the knee of the right internal capsule was found. The left side of the brain showed no gross lesion. The vessels at the base of the brain were very sclerotic.

MICROSCOPIC EXAMINATION. *Lumbar Region.* Sections from the lumbar region show a degeneration of both crossed pyramidal tracts, this being more marked on the right side. The direct pyramidal tracts are not degenerated.

Midthoracic and Cervical Regions. Sections from these parts of the cord show intense degeneration of each crossed pyramidal tract, more marked on the right side. The direct pyramidal tracts are not diseased. An area of slight degeneration is found in the posterior columns at the junction of the posteromedian septum with the posterior commissure.

The nerve cells in the lumbar and cervical regions, stained by the thionin method, do not take the color well. The cells are diffusely stained, the chromophilic elements are not sharply defined, and the pigmentation is very intense.

There is no evidence of meningitis at any part of the cord. The central canal in the lower cervical region is greatly dilated, forming a long slit extending from the neck of one posterior horn to that of the other. Here and there a very slight round-cell infiltration is seen around some of the vessels of the spinal cord.

Sections stained by the Marchi method show some recent degeneration in the right crossed pyramidal tract in the lumbar, thoracic, and cervical regions, but the degeneration in the left crossed pyramidal tract is of too long duration to show any degeneration by this method.

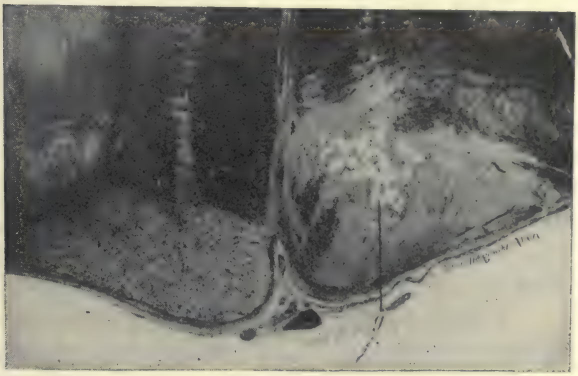

Fig. 6.—Section from the medulla oblongata (Case 2) stained by the Weigert hæmatoxylin stain, showing an area of softening in the left pyramid (*g*) and the adjacent lemniscus.

Medulla Oblongata. (Fig. 6.) The anterior pyramids are much degenerated. In the left anterior pyramid is an area of softening extending slightly into the lemniscus of the same side. This area of softening, when stained by a nuclear stain, consists of fatty, granular cells, packed tightly one against the other. There are no signs of meningitis.

Sections stained by the Marchi method from a lower level of the medulla oblongata show a very considerable recent degeneration in the left pyramid and a very slight recent degeneration in the right pyramid. The ventral portion of the left lemniscus is also partially degenerated. This is evidently the result of the area of softening previously described.

Pons. The pyramidal tracts are very much degenerated on each side.

Cerebral Peduncles. The middle portion of the foot of the right cerebral peduncle is degenerated. There is also a narrow band of degeneration in the innermost part of the foot on the same side. On the left side, by the Marchi method, there is recent degeneration in the foot of the cerebral peduncle, this area occupying approximately about one-half of the foot in its middle portion. An area of softening, as shown by the Marchi method, is found in the ventral part of the red nucleus, on the left side.

By the Marchi method several areas of softening are found in or near the knee of the left internal capsule and extending into the anterior portion of the posterior limb.

SUMMARY. Man, aged fifty-two years, not syphilitic. He had a number of attacks of hemiplegia, involving first the left side and later the right. Difficulty in speech and in swallowing became apparent after the second attack. All the extremities were weak, especially those on the left side. Sensation was normal. The patient was demented, had a bulbar speech, difficulty in chewing and swallowing, and dribbling of saliva. Macroscopically an area of softening was found in the knee of the right internal capsule. On the left side no gross lesion was apparent. Microscopically an area of softening was found in the medulla oblongata, this being of small size and confined to the left pyramid and a small part of the adjacent fibres of the left lemniscus.

Other areas of softening were found in the knee and in the anterior part of the posterior limb of the left internal capsule and in the ventral part of the red nucleus on the same side. Secondary degeneration of the pyramidal fibres was traced on both sides to the lumbar region of the spinal cord. The direct pyramidal tracts were not diseased. The degeneration of the median bundle of the foot of the right cerebral peduncle is evidently the result of the area of sclerosis in the knee of the right internal capsule. The degeneration of the middle part is probably due to a lesion higher in the brain, which was not found.

CASE III.—G. S., aged fifty-five years; baker; admitted to the nervous wards of the Philadelphia General Hospital December 23, 1901, in the service of Dr. Charles K. Mills. The patient was admitted from the Episcopal Hospital, where he had been for some time. The family history is unimportant; the patient had been an alcoholic. About one year before his admission to the Philadelphia General Hospital he had complained of general malaise, which was followed by numbness and impaired power of the left arm and leg. The paralysis was not complete, and there was no unconsciousness.

About one month before his admission, while sitting at a table eating, his employer noticed that the patient was staring very fixedly at him and was pointing to his mouth and endeavoring to signify that he could not talk. Examination the following day showed an inability of the man to speak, although he understood everything when spoken to, and he could write. The patient was right-handed. The pupils were equal and were normal in reaction to light. There was no extraocular palsy. The lower distribution of the left seventh nerve was paralyzed. The tongue was protruded to the left. The left arm and leg were somewhat paralyzed. Dysphagia was fairly well marked. The heart sounds were weak, the pulse was moderately full, and the face slightly flushed. The radial arteries were atheromatous. Sensation was retained in the pharynx, and the laryngeal reflexes on either side were present. The urine examination showed a large amount of albumin and tube casts. Examination of the throat by Dr. Gibb showed a paralysis of the muscles of the larynx, and the eye examination showed nothing abnormal.

The further notes, which were taken in the Episcopal Hospital, state that the patient improved considerably, and that the paresis in the left arm and leg became less. All the tendon reflexes of the upper and lower limbs were very much increased. Ankle clonus was obtained. The notes also state that the man could not fully protrude the tongue, and that dribbling of saliva was constantly present from both angles of the mouth, and that speech was indistinct, so that he could not be understood. A note is made also that the right side of the face seemed to be paralyzed.

On admission to the Philadelphia General Hospital the patient was in a delirious condition, and it was impossible to obtain a definite history. The eyes were staring, the mouth was open, and the tongue could not be protruded. The man could not understand anything that was said to him and had marked dysphagia, although he was able to swallow slowly. It was impossible to determine the state of the sensation, but the prick of a pin was evidently appreciated, as shown by the movements of the lower extremities. Both sides of the face were smooth and the mouth was not drawn in either direction. There was no absolute loss of power in any of the extremities. An examination was difficult. The lower limbs were œdematous, and there was marked cyanosis of the hands and feet. The notes, as taken by the resident physician, state that the patellar reflex was increased on the right and decreased on the left. The plantar reflex was present, Babinski's phenomenon being absent. The patient died December 26, 1901, three days after his admission to the hospital, after attempting to swallow milk. The autopsy showed congestion and œdema of the lungs, chronic interstitial nephritis, dilatation of the heart, and atheroma of the aorta.

MACROSCOPIC EXAMINATION. There is a scar in the substantia nigra of the left cerebral peduncle, as if from an old area of softening. At the uppermost portion of the right internal capsule there are three small cavities appearing as if they were areas of softening. One of these is situated in the anterior limb of the internal capsule near the knee; the other two are in the outer part of the lenticular nucleus, and involve the fibres passing to the posterior limb of the internal capsule.

MICROSCOPIC EXAMINATION. *Lumbar Region.* A distinct degeneration cannot be determined, because the material had been kept so long in formalin solution that the tissues do not take well the Weigert hæmatoxylin stain. So far as can be determined by the acid fuchsin stain, there is little or no degeneration of the white matter in this region. There is no round-cell infiltration of the pia. Most of the nerve cells of the anterior horns by the acid fuchsin stain are normal.

Thoracic Region. The sections from this region are not well stained, and it is better not to express an opinion as to the degeneration in the white matter. The bloodvessels are very much distended.

Cervical Region. Sections from this region are well stained and show a very distinct degeneration of each crossed pyramidal tract, more intense in the right. There is no degeneration of either direct pyramidal tract. Possibly there is a very slight degeneration of the columns of Goll, but this is somewhat doubtful. There is no round-cell infiltration of the pia. The nerve cells of the anterior horns stained by thionin are normal. The Marchi stain shows no recent degeneration, possibly because the sections were kept in formalin.

Medulla Oblongata. Both anterior pyramids are degenerated, but the left more so than the right. The intramedullary portion of each twelfth nerve seems to be normal, and the nuclei of these nerves, as stained by thionin, show nothing abnormal. The nucleus ambiguus is normal on each side. There is no round-cell infiltration of the pia. The bloodvessels of the medulla oblongata are much congested.

Pons. Both pyramidal tracts are partially diseased, the left more so.

Cerebral Peduncles. The median and adjacent portion of the foot of the right cerebral peduncle is degenerated. On the left side there is an area of degeneration in the substantia nigra, and in the adjacent middle portion of the foot of the cerebral peduncle.

Sections through one of the cavities found near the knee of the right internal capsule, stained by the nuclear stain, show softening of the tissues.

Summary. Man, aged fifty-five years; not syphilitic. He had several attacks of hemiplegia, which were confined to the left side. The notes did not give a distinct history of a right hemiplegia, but the examination showed a weakness of all the extremities, more so, however, on the left side. There was difficulty in speech and in chewing and swallowing. Saliva dribbled from the mouth. The mentality was poor. Macroscopic examination showed several areas of softening on the right side of the brain, these being situated in the anterior limb of the internal capsule near the knee, and in the outer part of the lenticular nucleus and extending above this nucleus, involving the fibres passing to the posterior limb of the internal capsule. On the left side there was a scar of an old area of softening in the substantia nigra of the left cerebral peduncle.

Microscopically areas of softening were found near the knee of the right internal capsule. The median and adjacent part of the foot of the right cerebral peduncle was degenerated. The secondary degeneration of the motor fibres of the right side, though not intense, was traced to the crossed pyramidal tract of the cervical region of the spinal cord. On the left side there was an area of softening in the substantia nigra of the cerebral peduncle. The middle portion of the foot was degenerated. The secondary degeneration of the motor fibres was traced to the cervical region.

Case IV.—C. M., aged fifty-one years; painter; admitted to the nervous wards of the Philadelphia General Hospital June 21, 1904. The family and past histories are negative. He has never had lead poisoning, but drank alcohol and smoked freely. The first attack of right hemiplegia occurred about a year before admission; the second, on the left side, five months after the first. Examination by Dr. Spiller, June 27, 1904, was as follows:

"The patient is not very intelligent. He has much difficulty in speaking, the speech being bulbar in character, and it is difficult to understand him. He does not seem to be aphasic in any way, and he understands what is said to him. He has much difficulty in swallowing fluid, chokes exceedingly, and cannot swallow a piece of dry bread. There is a distinct weakness of the lower distribution of the left seventh nerve, and he does not seem to be able to wrinkle his forehead well on either side. The masseter contracts firmly on each side. The tongue protrudes distinctly to the left, but is not wasted and shows no fibrillary tremors. Sensation for pain is normal over the face.

"The inward movement of the left eyeball is not perfect, and there is also some restriction in the inward movement of the right eyeball. He is said to have had this condition since infancy. The pupils are equal and react normally to light and in accommodation.

"The upper limbs are not distinctly weak. All the tendon reflexes are increased and equally so. Sensation is normal. There is no atrophy. Both lower limbs are distinctly weaker than the upper limbs. There is no atrophy, and sensation for pain is preserved. All the tendon reflexes are increased. Babinski's reflex is present on the right side and not on the left."

I had the opportunity of studying this patient many times before his death, which was caused by the development of a uræmic condition due to a chronic nephritis. Unfortunately, we were unable to obtain an autopsy.

Case V.—H. T., aged sixty-two years; loomfixer; admitted to the nervous wards of the Philadelphia General Hospital May 3, 1904. The family history is negative. There is no venereal history, but the patient was alcoholic. Two years before admission to the hospital the patient had a stroke, which came on slowly "like rheumatism." The patient states that he was paralyzed only in his left leg and that the left arm was not much affected. There was disturbance of speech at that time. One week before his admission he had another stroke, this time being unconscious, the paralysis involving the right side, and the leg more than the arm.

There is a slight ptosis of both eyelids, more marked upon the right side. The pupils are regular, the right being slightly larger than the left. The reaction to light and distance is normal. Convergence is normal. The eyelids can be tightly closed. The tongue is protruded slightly to the right, and movements can be performed in all directions, but are rather feeble.

The patient can understand what is said to him and can answer questions, but the speech is distinctly bulbar in character, being dull, monotonous and indistinct. The patient has the typical facies of pseudobulbar paralysis. Impulsive laughing and crying are present. He is not very intelligent, although he is able to obey commands.

All the extremities are weak, the lower more so. The tendon reflexes, both in the upper and the lower limbs, are much increased, and about equally so. The Babinski reflex is obtained on both sides, Sensation is normal. There is no apparent atrophy of any of the extremities. The gait is slow, somewhat spastic, and the steps are short.

Case VI.—H. P., aged fifty-one years; admitted to the nervous wards of the Philadelphia General Hospital October 10, 1904. Parents died of old age. Has had three children, one having epilepsy. No venereal history, but he used alcohol moderately.

Three years ago the patient had a right-sided hemiplegia, which diminished considerably. On April 28, 1903, he awoke one morning and found the left side of the body paralyzed. Since that time he has had several slight strokes, all of which involved the left side. He had difficulty in talking after his first attack, but this soon disappeared, and it was not until after his second stroke that his impairment of speech became apparent. He presents the typical appearance of pseudobulbar paralysis. The face is smooth and expressionless; the speech is thick, monotonous, and indistinct; saliva dribbles from the mouth, he is unable to swallow well, and he chokes occasionally. Forced laughing and crying are present.

The pupils are round, the left being slightly larger than the right, both reacting to light and in accommodation. Extraocular movements are normal. He cannot close either eyelid as well as he should. He can elevate either corner of the mouth nearly normally. The tongue is protruded straight, but the movement is weak. All the extremities are weaker than normal, the right more so. All the tendon reflexes, both in the upper and lower limbs, are increased, this increase being more marked on the right side. The Babinski reflex is present on both sides. There is hyperæsthesia for touch and pain in the right upper and lower limbs, this being more marked in the distal parts. There is no bladder or rectal disturbance.

The mental condition is poor, the patient being demented. The urinary examination shows a moderate amount of albumin and hyaline and granular casts.

SUMMARY. The clinical cases without necropsy do not present any unusual symptoms. All the patients had history of more than one hemiplegic attack, the extremities were weak, the reflexes were increased, there was difficulty in articulation, deglutition and mastication, and dribbling of saliva was always present. The mentality was impaired in every case. In Case VI. there was some atrophy in the distal portions of the upper and lower extremities of one side, and hyperæsthesia for touch and pain upon the same side.

Joffroy, in 1872, first described the symptom-complex of pseudobulbar palsy under the title of "Paralysie labio-glosso-laryngée d'origine cérébrale," and it remained for Lépine, in 1877, to give the disease the term of pseudobulbar paralysis. Among the German writers, Berger, Jolly, and Kirchhoff were among the first to describe this symptom-complex.

Pseudobulbar paralysis generally appears about middle life, and is more frequent in the male sex. There is at first an attack of hemiplegia which, in a short time, is followed by one or more similar attacks involving either the same or the opposite side. As a rule, the loss of power in the extremities is not very marked, and we may have the extremities of one side weaker than those of the other, or there may be paresis of one side only. The symptoms of pseudobulbar palsy may be present without any weakness of the extremities. Disturbances in speech, in chewing, and in swallowing may appear directly after the first hemiplegic attack, or, as is more often the case, not until after one or more of the subsequent attacks. As a rule, a partial return of power in the extremities is found, but, unlike what we have in hemiplegia, the return of power in the upper extremity is much greater than in the lower extremity. The limbs are spastic, but not very much so, and contractures are not very frequent, but when they do occur they differ from the usual contractures seen in hemiplegia. The greater involvement of the lower limbs and the difference in the contractures can be explained by the location of the lesions which are numerous, differing from the unilateral lesion causing hemiplegia.

All the tendon reflexes of the extremities are increased. Atrophy of the limbs is unusual, but it does occur, as in one of my cases (Case VI.). When present, the atrophy is not very marked, and involves principally the distal parts. This can readily be explained, for it has been shown by Marinesco, myself, and others that atrophy in hemiplegia is a more or less constant symptom of the affection. Disturbances of sensation are unusual. In one of my cases (Case VI.) there was a hyperæsthesia for touch and pain, this being also more marked in the distal parts. The electrical reactions are not altered.

Vasomotor phenomena, as blueness of the face and of the extremities, have been noted. It is not unusual to see the face become red during attacks of impulsive laughing and crying. Variations in the pulse have also been noted. Oppenheim and Siemerling recorded a case where during the attacks of impulsive laughing and crying the face became livid, the breathing, especially in expiration, ceased for a very short time, and the pulse was small and arrhythmic. Disturbances in respiration are not unusual. I have shown that in hemiplegics the *respiratory excursion* on the paralyzed side is less both in quiet and in forced respiration. Attacks of coughing and choking are common. Cheyne-Stokes breathing has also been noted. Respiratory cramp has been mentioned by Jolly.

The facial nerve may be paralyzed on each side or only on one. In some cases it is not all involved. The upper distribution of the facial nerve, as a rule, is not paretic. The patient, therefore, especially as the disease progresses, presents what is known as the pseudobulbar facies. The cheeks droop, sometimes on both sides or only on one; the face is smooth and expressionless; the eyes are staring, the lids being wide open; the lips are apart, the lower one sometimes drooping, and saliva constantly dribbles from the mouth.

The speech, as mentioned above, may be disturbed after the first attack or not until the subsequent attacks, and there may be only a very slight, hardly perceptible disturbance, or complete aphonia. As a rule, the speech is dull and monotonous, and, as Brissaud has so well expressed it, the song of the speech is lost, and it is difficult for the patient to make himself understood.

Mastication is disturbed from a very slight degree to a total inability of the patient to chew. This is caused either by a paresis of the masseter, temporal, and the pterygoid muscles on one or both sides, or, what is more common, it is due to the paresis of the lips, tongue, and palatal muscles.

The tongue is nearly always partially paralyzed. The movements are not well performed, and the tongue cannot be protruded fully; but there are no fibrillary tremors or atrophy. Fibrillary tremors, however, have been reported in a very few instances.

The palatal muscles either on one or both sides may be paretic, and the palatal reflex is generally lost. The pharyngeal and laryngeal muscles may also be paralyzed, and the pharyngeal reflex lost. Disturbances in phonation are common, as are disturbances in deglutition. This disturbance may be partial or may be total, so that the patient is unable to swallow anything but liquids, and even then the food regurgitates often through the mouth and nose. Choking is very common.

Impulsive laughing and crying are very common symptoms of pseudobulbar palsy, although they are not present in every case. They are generally late manifestations. They are entirely beyond the control of the patient, coming on at any time, and may be brought on by the slightest irritation, or may occur without it. In some cases either impulsive laughing or crying is present alone, though it has seemed to me that crying is more often present alone. Crying may terminate in laughing, or *vice versa*. These acts involve a complicated musculature, and it is noticeable that when they do occur, the whole musculature is involved, differing from the ordinary laughing and crying, in which only a part or all the muscles concerned in this act may be involved. It is sometimes possible for the patient in the mild cases to voluntarily cry or laugh, but this will invariably bring on a furtherance of these acts. This forced laughter or crying is a spasmodic condition of the muscles concerned in the act. The secretion of tears is unusual, but I have seen the eyes fill up with tears in a patient suffering from multiple sclerosis with impulsive laughing and crying.

Saliva constantly dribbles from each corner of the mouth, and it is a question whether the secretion of saliva is not abnormally increased. Cases are on record, however, where the secretion of saliva was diminished, leaving the tongue and mouth in a dry condition.

Ocular symptoms are rare. The pupils have been found unequal in size, as in one of my cases, and unilateral myosis with pupillary rigidity has been recorded by Knotz. Bilateral pupillary rigidity has also been recorded. Optic neuritis and atrophy are rarely present, but were in a case of Marshall and Jacob, in which there were blindness and also nystagmus. The ocular muscles are rarely involved. Hartman recorded a case with slight bilateral ptosis and loss of convergence. Wernicke was the first to point out that in bilateral supranuclear lesions there may be what is called external pseudo-ophthalmoplegia. Oppenheim and Hartman record similar cases. In Oppenheim's case the patient could not voluntarily look to either side, but could look upward and downward. He could follow the finger, however, in any direction. In another one of his cases the patient could not look toward the right, but could toward the left. This inability to perform voluntary associated movements when power to perform involuntary associated movements is retained, was first pointed out by Rose, and seems characteristic of this disease.

Involuntary movements or contractions may occur in the course of pseudobulbar paralysis, not only in the extremities, but also in the distribution of the fifth, seventh, and twelfth nerves, and in the muscles of the neck. Oppenheim records an unusual case with bilateral choreic or athetoid movements. This, however, occurred in a case of infantile pseudobulbar palsy, for it is unusual for these movements to occur in the adult, even in an ordinary hemiplegia. Within a few days I had occasion to see with Dr. J. W. McConnell a woman, aged thirty-seven years, who had a questionable right-sided stroke one year ago, and who, three days before I saw her, had another stroke, this time on the left side. She had hardly any perceptible loss of power in the extremities of either side, but had left hemiataxia, hemianæsthesia, and hemianopsia, and continuous, irregular rotatory movements of the left upper limb, which reminded one of athetoid movements. This very interesting case was fully reported by Dr. McConnell.

The muscles of the neck are not usually involved in involuntary spasm. The face and tongue are rarely the seat of contractions. Bamberger records tonic and clonic tremor of the face. Tic is rare. Lépine records trismus. In a patient I have studied in the care of Dr. C. S. Potts there is a constant retraction of the risorius muscle of each side, causing the patient to have the appearance of a continuous grin. Oppenheim records spasm of the masseter and temporal muscles, causing tremor of the lower jaw and chattering and grinding of the teeth, this interfering with any use of the jaws. In Bamberger's case there was also spasm of one pterygoid muscle.

The intelligence of the patient is usually affected early in the disease. It is not unusual to have slight failing of mentality after an ordinary attack of hemiplegia, so it is not remarkable that the mental functions are much altered on account of the severe multiple lesions of pseudobulbar paralysis. There may be mental apathy, melancholia, or violent maniacal outbreaks—in fact, any of the mental disturbances common in the aged. As a rule, the patient is quiet and stupid, does not bother himself with anything, and prefers to be let alone.

The symptoms are progressive, and the patient in a short time presents what is known as the typical picture of pseudobulbar paralysis. The French authors have not unjustly likened the late appearance of this condition to that of paralysis agitans. We have in both the rigid immobile facies, and we may have even in paralysis agitans involuntary laughing and crying. When walking the patient stoops over, the steps are short, the knees are not brought up high from the ground, and the whole attitude of the patient is stiff. In more severe cases, however, the patient is confined to his chair or bed. Death usually occurs in the course of a few years, due to some intercurrent disease, to another hemorrhage, or, what is not unusual, from choking while attempting to swallow food.

PATHOLOGY. It must be understood that pseudobulbar palsy is not a definite disease, but a symptom-complex brought on by a variety of pathological conditions, chief of which is arteriosclerosis. The basal arteries, as the vertebral and basilar, are always found in a sclerotic condition, and their prolongations into the brain tissues are, of course, similarly involved. We can readily see, therefore, that the basis of the disorder is an alteration of the bloodvessel walls, which causes miliary aneurysms or thrombosis, resulting in hemorrhages or areas of softening. These hemorrhages and areas of softening may be found anywhere in the brain substance, and may be of large size, or so small that only careful microscopic examination will reveal them.

Probably from anatomical reasons they are more frequently in the central ganglia than in the cortex, subcortex, pons, or medulla oblongata. The majority of writers upon this subject insist upon the preponderance of the lesions in the central ganglia, especially in the putamen or near the lenticular nucleus. Others insist upon the majority of the lesions being in the projection fibres coming from the Rolandic operculum, while still others, as Oppenheim and Siemerling, believe that we have not a pure cerebral type of the disease, but that a careful microscopic examination will always show microscopic areas of softening in the pons and medulla oblongata.

Urstein's statistics are interesting. Of the cases with macroscopic and microscopic examination forty had bilateral lesions in the brain. In fourteen the lesions were unilateral. I have examined into the latter number and could verify only nine such reported cases. In twenty-three instances the pons and medulla oblongata were macroscopically and microscopically normal. In thirteen there were, in addition, lesions of the pons, the medulla oblongata being normal. In two cases there were lesions of the pons only, the brain and the medulla oblongata being normal.

In my cases, diffuse lesions were found throughout the brain and the brain stem. In Case I. on one side, large areas of softening were found in the lenticular nucleus and internal capsule (Fig. 9), and on the other side, a lesion in the knee of the internal capsule (Fig. 10). A syphilitic infiltration of the pia was found throughout the brain and cord, and the bloodvessels were sclerotic. In Case II. an old scar was found in the knee of one internal capsule, and on the other side an area of softening in the anterior pyramid in the medulla oblongata (Fig. 11). A careful search for the cause of the primary motor degeneration of the same side revealed other areas of softening in the knee of the internal capsule and in the region of the red nucleus. The bloodvessels were sclerotic throughout. In Case III. there were areas of softening on one side in the corona radiata, cutting off the fibres going to the posterior limb of the internal capsule and on the other side there was an area of softening in the substantia nigra.

The study of these cases has impressed upon me the necessity for careful and thorough microscopic examination, for it is evidently unsafe to express opinions regarding the extent of the lesions found in this disorder unless all parts of the brain and brain stem have been thoroughly examined. Oppenheim and Siemerling were the first to point this out, and, I think, rightfully insisted upon a careful examination of the medulla oblongata and pons for lesions in these areas, but I also believe that this examination should extend to all parts of the brain. From the nature of the etiology of the symptom-complex—an arteriosclerosis of the cerebral vessels—we rightfully expect lesions in any part of the brain and brain stem.

This should be remembered in the cases where unilateral lesions were supposed to have caused symptoms of pseudobulbar paralysis. I have been able to collect nine such cases. In the early recorded instances before the Marchi method was used, examination was necessarily incomplete. In the later instances it is probable that a more complete examination of the cortex and other parts of the brain would have led to the finding of areas of softening, possibly minute. This is well demonstrated in Case II., where no explanation was found at first for the recent primary degeneration of the pyramidal fibres of one side, but further study by the Marchi method showed areas of softening in and near the knee of the internal capsule. It might be well to state here that the cortical cells, as shown in the condition of the Betz cells of the paracentral lobules, were not altered.

Recently Concetti recorded a case of pseudobulbar palsy in a boy, aged five years, in which at the autopsy the right middle cerebral artery was found thrombotic. According to him, in the blood supply of this artery are included the corticobulbar and the gangliobulbar centres and the crossed fibres from the opposite hemisphere. Therefore, the medulla oblongata was not only deprived of the influence of the injured, but also of the healthy side, through the involvement of the crossed fibres. In this instance the lesion, while anatomically unilateral, involved fibres from both sides of the brain.

Besides the hemorrhages, old apoplectic cysts, and areas of softening found with macroscopic and microscopic examination in the cortex, corona radiata, internal capsule, central ganglia, cerebral peduncles, pons, and medulla oblongata, ependymitis of the floor of the fourth ventricle and internal hydrocephalus have been described by Oppenheim and Siemerling. Jolly reported pseudobulbar symptoms in a case of multiple sclerosis, the areas of sclerosis being confined to the lenticular nucleus of each side. Pseudobulbar symptoms have also been reported as the result of endocarditis, this causing thrombosis of the cerebral vessels. Brauer and v. Halban found encephalitis, and Burr and McCarthy diffuse cortical pachymeningitis. In a case of Comte's there was, through an acquired syphilis, a sclerosis with a disappearance of the cellular elements in the operculum of each side. Pseudobulbar symptoms have also been recorded without anatomical lesion, and also as due to peripheral neuritis of cranial nerves and to hysteria. This will be discussed later.

Oppenheim recorded, in 1895, the first case of infantile pseudobulbar palsy. His report was rapidly followed by that of Bouchaud and others. In the few recorded cases with autopsy there has nearly always been found a congenital maldevelopment of the cortex. In Oppenheim's case there was bilateral microgyria in the Rolandic gyri and porencephalus on the left side in the mid-portion of the Rolandic gyri. In Bouchaud's case there was microgyria involving symmetrically the lower Rolandic convolutions.

Zahn reported an interesting case of congenital pseudobulbar palsy in which, microscopically, a primary hypoplasia of the corticobulbar tracts was found. Concetti's case of thrombosis of the mid-cerebral artery has been mentioned. In Marshall and Jacob's case bilateral tumors the size of a walnut were found. The tumor on one side was located in the lenticular nucleus and invaded the internal capsule and optic thalamus, and on the other side the tumor was in the optic thalamus and also encroached upon the internal capsule. The tumors consisted of caseated granulomata. There was also a tumor in one occipital lobe. There were no other symptoms of tuberculosis, and, curiously enough, none of the symptoms of brain tumor.

Conclusions concerning the Pathology. We can come to the conclusion, therefore, that pseudobulbar palsy in the adult is probably always caused by bilateral lesions, and that these lesions may be in the cortex, subcortex, internal capsule, central ganglia, as the caudate nucleus, optic thalamus, especially the lenticular nucleus, cerebral peduncles, pons, or medulla oblongata. These lesions consist either of areas of softening or hemorrhages which can be seen macroscopically, or may not be found unless a careful microscopic examination is made. The central ganglia, especially the lenticular nuclei, are more often involved than the other parts of the brain. It is impossible to make a pathological distinction between a pure cerebral and a cerebropontile type, unless a thorough microscopic examination has been made, and if so it will be found that there are diffuse lesions throughout the brain, and probably, also, in the medulla oblongata and pons. The cause of the whole symptom-complex is usually an arteriosclerosis of the bloodvessels of the brain.

It is probable that in the cases of pseudobulbar palsy reported as caused by unilateral lesion a more careful microscopic examination of the brain would have shown bilateral involvement.

In infantile pseudobulbar palsy the cause is mostly congenital maldevelopment, although cerebral thrombosis and bilateral tumors in the basal ganglia may cause the symptom-complex of pseudobulbar palsy.

Etiology. The symptoms of pseudobulbar palsy differ from those of the true Duchenne type of bulbar palsy, because the lesions in the former are above the nuclei of the different cranial nerves; therefore, a supranuclear lesion of the fibres coming to these nuclei, anywhere in their course or in their cortical centres, may cause the symptom-complex of pseudobulbar palsy.

It is taught that unilateral lesions of these bulbar fibres or of the cortical centres will not cause permanent symptoms, because the bulbar muscles have a bilateral cortical representation. We have, however, even in unilateral lesions, as in hemiplegia, some symptoms of bulbar involvement in the first few days following the attack. I am unwilling to believe that a unilateral lesion will not produce any permanent impairment of muscles having a bilateral cortical representation. It is not to my mind conceivable that an injured mechanism can so fully recover all of its functions from the influence of the remaining centre.

I have shown that in the respiratory movements of hemiplegics there is not an increase of respiratory movement due to a loss of cortical inhibition, as has been heretofore taught, following the investigation of Hughlings Jackson, but that there is an actual decrease in the respiratory movement of the paralyzed side, both in quiet and in forced respiration. Again, the other bilateral mechanisms, as shown in articulation, phonation, and deglutition, are always somewhat disturbed in hemiplegia. In other words, a unilateral destruction of the bulbar fibres, or their cortical centres, will always disturb the functions involved.

It can be readily seen, therefore, that a bilateral disturbance of muscles dependent upon bilateral cortical centres will produce severe symptoms in the acts of articulation, mastication, phonation, and deglutition—*i. e.*, will cause the symptom of pseudobulbar paralysis.

Bulbar symptoms have been reported as due to peripheral bulbar neuritis. Such cases have been noted by Kast and Eisenlohr. Again, a form of bulbar palsy has been noted by Hori and Schlesinger, in which there were besides the symptoms of an acute hemorrhagic poliomyelitis superior, the typical findings of that disease. Again, this symptom-complex has been recorded as due to hysteria. Schlesinger also recorded such symptoms in a case of tumor involving the uppermost part of the spinal cord. Spiller and I reported bulbar symptoms in a case of vertebral carcinomata. While we have in all of these instances bulbar symptoms, it is inadvisable to liken these to the pseudobulbar phenomena due to multiple lesions of the brain.

ANATOMICAL FINDINGS. In Case I. on the right side there was found an area of degeneration in the internal capsule occupying approximately about one-fifth or one-sixth of the entire area of the posterior limb. The middle part of the foot of the right cerebral peduncle was found degenerated, this degeneration occupying the whole extent of the right pyramid in the medulla oblongata, while in the spinal cord the right direct pyramidal tract was more intensely degenerated than the left crossed pyramidal tract, both in the cervical and the midthoracic regions, as shown by the Weigert hæmatoxylin and Marchi methods. In the cervical cord the degeneration extended from the periphery of the cord back to the white commissure. In the lumbar cord the left crossed pyramidal tract was still diseased, the right direct pyramidal tract being normal.

These findings are interesting. The degeneration of the motor tracts in the internal capsule was caused by a higher lesion, which was not found.

The degeneration of the direct pyramidal tract was greater in intensity than that of the crossed pyramidal tract, and this can be readily explained by the well-known variability of the motor decussation in the bulb.

On the left side, by the Weigert method, an area of softening was found directly in the knee of the internal capsule. In the peduncle, the innermost part of the foot was found degenerated. We have here, therefore, a degeneration of the median bundle of the peduncle as a result of an area of softening in the knee of the internal capsule.

Dejerine, in three cases, studied by the Weigert method the secondary degeneration from lesions limited to the Rolandic operculum and the adjacent part of the frontal operculum, and traced this degeneration to the knee of the internal capsule and into the median bundle of the peduncles. Zacher believes that this part of the crusta is chiefly connected with the insula, while Flechsig and Wernicke think that this bundle comes from the frontal lobe. v. Monakow is also in favor of the latter view. In Spiller's case the internal bundle of the peduncle was well stained, while the operculum was degenerated, and he concludes that the origin of a portion of this bundle is anterior to the Rolandic fissure. In K. Schaffer's case the whole frontal lobe was destroyed, causing a degeneration of the anterior limb of the internal capsule and of the median part of the crusta.

More interesting, still, is the further degeneration of the motor tracts in Case I. In the pons one or two bundles on the left side near the raphe and in the ventral part were degenerated. In the medulla oblongata the outer part of the left pyramid was slightly degenerated. In the cervical and thoracic regions of the cord the left direct pyramidal tracts was degenerated to a considerable degree, not so intensely, however, as the direct pyramidal tract of the other side. The right crossed pyramidal tract was normal, both by the Weigert and Marchi methods.

We have here, therefore, a slight degeneration of the motor tracts which was traced through the pons and medulla oblongata and into the direct pyramidal tracts of the cervical and dorsal regions. The degeneration of the right direct pyramidal tract is evidently a result of the area of softening found in the knee of the left internal capsule, and is a continuation of the degeneration of the median bundle of the foot of the cerebral peduncle. Such a finding is entirely at variance with our knowledge of the course of this tract, but we are compelled to come to the conclusion that a degeneration of the median bundle of the foot of the central peduncle may cause a degeneration of the direct pyramidal tract. This, however, cannot be definitely accepted, without further anatomical or pathological proof.

The bilateral degeneration of the direct pyramidal tract with only a partial degeneration of one crossed pyramidal tract from lesions in the brain substance seems to be a contradiction to the views recently advanced by Marie and Guillain. These authors assume that the direct pyramidal tracts consist of two sets of fibres, one from the cortex and the other from the mesencephalon. According to them, a cortical lesion will produce only a very slight degeneration of the direct pyramidal tract, while a lesion limited to the peduncles or pons will produce an extensive degeneration occupying the whole extent of the direct pyramidal tract.

Ugolotti first criticised these findings, and expressed the opinion that the degeneration in this tract did not depend upon the place of the lesion. M. and Mme. Dejerine have criticised more severely Marie and Guillain's views, and have stated that their conclusions were not warranted by facts, and that the variation in the degenerations of the direct pyramidal tracts were due to the well-known variability of the motor decussation in the bulb.

Marie and Guillain in a later paper reassert their original views. That their views are not born out by facts my cases prove, for we have on one side a lesion in the internal capsule in which the resulting degeneration in the cord is greater in the corresponding direct pyramidal tract, and on the opposite side the lesion was confined to the knee of the internal capsule, while the degeneration was strictly limited to the direct pyramidal tract. This degeneration in the cervical cord was traced by the Weigert and the Marchi methods.

I have examined in this connection still another case in which there was an area of softening limited to the pyramidal fibres of one side of the pons, except that it slightly invaded the median fillet of the same side. The brain was normal. The degeneration in the cervical cord, both by the Weigert and the Marchi methods, was strictly confined to the crossed pyramidal tracts, the direct pyramidal tracts being entirely normal.

I am indebted to Dr. C. K. Mills and to Dr. C. S. Potts for the material of two of these cases, and to my chief, Dr. William G. Spiller, for the material of the third and the privilege of reporting the clinical cases.

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MUSCULAR ATROPHY, DEGENERATION OF THE TRIGEMINUS NERVE AND OF THE LATERAL COLUMNS, AND ANÆMIC CHANGES IN THE SPINAL CORD OCCURRING IN TABES DORSALIS.

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Tabes dorsalis is an organic disease of the nervous system which clinically and pathologically has been extensively discussed, but which, despite this fact, presents many symptoms and pathological aspects still meriting our attention.

A large number of observations have been made on the occurrence of muscular atrophy in tabes, and this condition does not appear to be rare. The statistics of Dejerine[1] and of Joseph Collins[2] show that from 15 per cent. to 20 per cent. of all cases of tabes present this complication, which is, in part at least, responsible for the deformity of the foot known as the tabetic club-foot.

While the lower limbs are the most frequently affected, they are by no means always involved, and, in fact, the atrophy may have almost any distribution, frequently attacking the upper limbs, beginning in the hand, or may even present the scapulohumeral type, as observed by Dejerine.[3] Belonging, also, to this category are those cases in which there has been observed an atrophy of muscles which have their centre of innervation in the bulbar nuclei, as the muscles of the tongue, which have been reported unilaterally atrophied in the cases of Raymond and Artaud,[4] Koch and Marie,[5] and others.

Through the generosity of Dr. William G. Spiller I am enabled to report, with pathological findings, a case of tabes dorsalis from his service at the Philadelphia General Hospital, which presented, in addition to a marked atrophy of both lower and to a slight extent of the upper extremities, an intense anæmia, and trophic disturbances in the distribution of the fifth nerve. The notes, taken on July 16, 1902, are as follows:

William J. H., aged forty-seven years. Denies syphilitic infection. Presents the diagnostic symptoms of tabes which began with ataxia eight years ago.

On examination the patient shows ataxic station and gait, increased by closing the eyes; lost knee-jerks and bicep-jerks; incontinence of urine; Argyll-Robertson pupils, the left being oval and slightly larger than the right; and delayed sensation below a line drawn through the umbilicus. The feet are deformed, presenting the typical tabetic club-foot, or "pied-bot tabétique" of the French writers. The muscles of the lower extremities are extremely atrophied and are very weak. The muscles of the arms are wasted, but they are not weak. He has two indolent ulcers on the right foot. He has lost several teeth, which he says he extracted without pain. The patient presented the appearance of extreme anæmia, which was verified by a blood examination.

Death was due to œdema of the lungs, and at autopsy he was found to have a chronic interstitial nephritis and a psoas abscess, with erosion of the fourth and fifth lumbar vertebræ. The brain and cord were removed and preserved for histological examination.

Microscopically the changes in the posterior columns of the spinal cord are those of advanced tabes. In the lumbar region the degeneration involves the whole of the posterior columns, except the cornu-commissural zone. As is usual in tabes, however, many fibres have escaped. In the middle thoracic region the posterior columns and posterior roots are intensely degenerated. In the cervical region the posterior roots are much affected.

The intervening zone of v. Bechterew, to which attention has recently been drawn by Reichardt,[6] is certainly degenerated throughout the cervical region as high as the motor decussation.

In a section obtained from the cervical region a mass of swollen axis cylinders is found in one anterior column, near the anterior commissure. The axis cylinders here are much swollen, but there is no round-cell infiltration in this area. A similar group of swollen axis cylinders is seen in a section from the thoracic region. In this section they are found near the periphery of the cord, in the region of Gowers' column, and here, likewise, there is no round-cell infiltration. The swollen axis cylinders are found in a loose reticular tissue, and spaces of considerable size are found about them, even about those only moderately swollen. These changes are very much like those occurring in the spinal cord in cases of anæmia. They are not inflammatory in type, as shown by the absence of round-cell infiltration, and they do not appear to be in the neighborhood of blood-vessels.

A very slight round-cell infiltration is seen in the pia of the cervical and thoracic region.

Sections of the right fifth nerve in the extrapontile portion appear partially degenerated when stained by the Weigert hæmatoxylin method. The descending spinal root of the fifth nerve in the medulla oblongata appears to be normal, so that if the fifth nerves were degenerated they could have been only slightly so. The degeneration in such a case might be less distinct in the lower part of the spinal root.

Many of the nerve cells in the anterior horns in the lumbar region are diseased. Some are nearly or fully normal. The changes consist of a chromatolysis which involves often the whole cell and cannot be said to be distinctly central or distinctly peripheral. Frequently the cells appear as masses of fine purple granules, when stained with thionin. In some the nucleus is displaced to the periphery, and often the dendritic processes have completely disappeared. The changes are not those described by Marinesco as secondary to degeneration of peripheral fibres, his "reaction at distance," and if any importance is to be attributed to this distinction the degeneration of the anterior horn cells in this case cannot be regarded as secondary to an alteration of the nerve fibres.

To summarize the important clinical features of this case: we have the diagnostic symptoms of advanced tabes complicated by muscular atrophy, most marked in the lower extremities; an intense anæmia, possibly due to a suppurative process; the psoas abscess, and trophic disturbances in the distribution of the fifth nerve, as shown by the painless falling out of the teeth. Pathologically the case shows the lesions of advanced tabes dorsalis in the posterior columns of the spinal cord, and in the posterior roots; marked degenerative changes in the nerve cells of the anterior horns of the spinal cord, viz., chromatolysis, displacement of the nucleus, and loss of dendritic processes; beginning changes in the cord produced by the anæmia, shown by the swollen axis cylinders in the anterior columns, and partial degeneration of the fifth nerves.

Another case of tabes occurring in the service of Dr. Spiller, at the Philadelphia General Hospital, presents interesting pathological features. In this case there was no atrophy, and in that respect it is valuable for comparison with the first case (W. J. H.).

The clinical notes, taken in 1901, are as follows:

John A., aged fifty-five years. His family history and previous medical history were negative. He presents the symptoms of tabes dorsalis, which is of about six years' duration. The ataxia is extreme. He cannot walk or stand unsupported. His pupils do not react to light, and but feebly in accommodation. The right pupil is dilated. There is "ataxia" on lateral deviation of the eyes. His tongue presents no tremor or atrophy. The arms are slightly ataxic, but there is no tremor or atrophy. There is no atrophy of the legs. The knees can be slightly hyperextended. The tendon reflexes are normal in the arms, but absent in the legs. He complains of lancinating pains in the legs. Pain and thermal sensations are lost below the middle of the leg, except in the soles of the feet. Sensation to touch is normal in all parts of the body.

Death was due to œdema of the lungs and chronic interstitial nephritis. At the autopsy, conducted by Dr. R. M. Pearce, the following notes were made: The body is well developed and well nourished. The dura mater is more or less thickened throughout the entire extent of the spinal cord, but most markedly about 5 cm. above the termination of the cord, extending upward for a distance of 10 cm. In this area the dura is not only thickened but adherent to the pia, which is also thickened, forming a dense, cord-like mass containing calcareous material. The cord throughout this area is much smaller than normal. On section the entire circumference of the cord is seen to be diminished. The posterior columns, and to a slight extent the posterolateral, are smooth, glistening, and translucent, as compared with the opaque, white, anterior half of the cord. This degeneration is visible throughout the entire length of the cord.

Sections from the spinal cord in the thoracic and lumbar regions, microscopically examined, show intense lesions of tabes in the posterior columns and posterior roots. In the cervical region the posterior roots are only partially degenerated. The intervening zone of v. Bechterew is degenerated throughout the entire cervical region of the cord, up to the motor decussation.

In the lumbar region (Fig. 1) a zone of degeneration is found in the posterior part of each lateral column, not extending as far as the posterior horns. It is irregular in shape, and does not correspond closely to the configuration of any known tract. This lesion was probably caused by some interference with the vascular supply, most likely by the adhesion of the dura, as described in the post-mortem record. In the midthoracic region (Fig. 2) the degeneration of the lateral columns at the periphery is intense on each side, but it is irregular in distribution, and apparently vascular in origin. In the cervical region the peripheral degeneration is still present in the lateral columns, but is more in the area of the columns of Gowers than in the direct cerebellar tracts.

There is no round-cell infiltration of the pia mater of the spinal cord at any level.

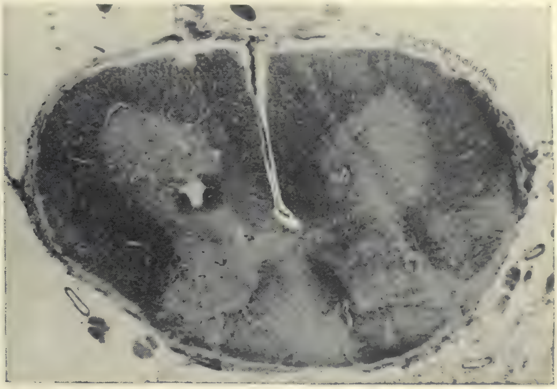

Fig. 1.—Section from the lumbar region of the spinal cord in a case of tabes stained by the Weigert hæmatoxylin method. The posterior columns and each lateral column, especially the left, are degenerated. The section is reversed in the photograph.

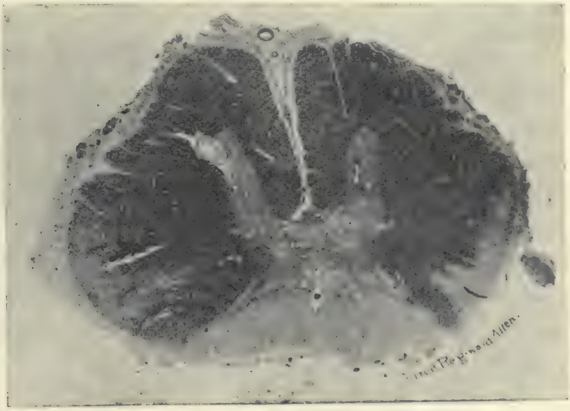

Fig. 2.—Section from the midthoracic region, showing a similar degeneration to that in the lumbar region (Fig. 1).

The walls of many of the bloodvessels are much thickened, and the intima of the anterior spinal artery in the lumbar region is much proliferated.

The nerve cells of the anterior horns of the spinal cord are highly pigmented, but otherwise are not much altered.

The most interesting pathological finding in this case is the irregular degeneration of the lateral columns, which was probably vascular in origin. Another important feature of the case was the correspondence of the healthy nerve cells of the anterior horns with the unaltered musculature.

The muscular atrophy occurring in tabes exhibits clinically a diversity of form, as well as distribution. So marked are these different aspects in some cases that P. Marie[7] divides all cases into two classes. In his first class are those in which the amyotrophy comes on late in the disease, is symmetrical, and is not generally accompanied by fibrillary contractions. Included in the second class are those cases in which the atrophy begins early, is less frequently symmetrical, and is often accompanied by fibrillary contractions and the reaction of degeneration.

It seems to me that this distinction is somewhat artificial. There are cases reported of all gradations between the two types, and in a disease such as tabes dorsalis, which clinically can be made up of so many different symptom-complexes or types, the cervicobulbar, the ocular, etc., it is much more logical to suppose that the amyotrophies are essentially the same in all cases, differing only in the location and virulence of the disease process.

The most frequent location of the atrophy is in the lower limbs, first affecting the peroneal group of muscles, and is slowly progressive, producing the tabetic club-foot, particularly studied by Joffroy,[8] and present in my first case. According to Marie, the feet are in a position of permanent exaggerated extension, and turned slightly inward, producing, in effect, a talipes equino-varus, and is described as such by Bornstein[9] and others.

Bornstein agrees with Marie in his division into two classes of atrophy, yet in both of his cases the atrophy came on late in the disease and attacked the lower limbs, but in the first case it was asymmetrical, and in the second showed the reaction of degeneration. The toes are flexed markedly in the tabetic club-foot, but the foot can generally be freely moved laterally. The palpation of the muscles shows flaccidity. "It is not the club-foot of contracture, of infantile spinal palsy, nor is it of an osseous nature, but a club-foot of flaccidity, an atonic club-foot" (P. Marie).[10] Rarely there are cases of atrophy without deformity (Jos. Collins).[11]

Atrophy in tabes is by no means confined to the lower limbs, but may be localized almost anywhere, in the area of innervation of the bulbar nuclei, in the upper extremities, or in the muscles of the trunk, particularly the scapular region. In some cases the atrophy is slowly progressive, similar to that occurring in the Aran-Duchenne type of progressive muscular atrophy.

An examination of the reported cases shows the wide range.

Friedreich[12] describes a case with atrophy of the hypoglossus. Raymond and Artaud,[13] in a case with hemiatrophy of the tongue and slight atrophy of the right thenar eminence, found pathologically atrophy of the hypoglossus nucleus and nerve, and degeneration of the inner group of cells in the anterior horns of the cervical region. The motor trigeminal and vagus nuclei showed changes. Eisenlohr[14] reported a case of tabes with symmetrical atrophy of the tongue, a slight interference with the movements of the lips, and palatal muscles, paralysis of the postcricoarytenoid muscle, and of the vocal cords. Anatomically, there was found a degeneration of the posterior columns and posterior roots; atrophy of both vagus, glossopharyngeus, accessorius, and hypoglossal nerves, and of both recurrent nerves, and atrophy of many cells in the tenth, eleventh, and twelfth bulbar nuclei. Koch and Marie observed a case with hemiatrophy of the tongue and atrophy of the hypoglossus nucleus of that side. Westphal,[15] Demange,[16] Kahlor,[17] Landouzy and Dejerine[18] have also found lesions in the bulbar nuclei in tabes. Leyden, in his paper as early as 1863, "Die graue Degeneration der Ruckenmarkstränge," described three cases with atrophy, in two, in the lower extremities and in a third beginning in the small muscles of the hand. The anatomical findings in these cases are not given.

A study of the literature on this complication of tabes, with great detail, is given in Dejerine's work on the pathological anatomy of muscular atrophy in tabes.[19] More recently papers on this subject have been written by Goldscheider[20] (1891), Jolly[21] (1891), Ufen[22] (1896), Wagner[23] (1896), Chretien and Thomas[24] (1898), Müry[25] (1901), Raymond and Phillip[26] (1902), E. W. Taylor[27] (1902), Simin Kiwi[28] (1902), Freidrich Rein[29] (1903), Köster[30] (1903), and others.

The pathological condition responsible for the muscular atrophy occurring in tabes is not definitely determined. Some authors, notably Dejerine, have considered it due to a motor neuritis; others, following Charcot, have considered it as of central origin—*i. e.*, due to changes in the nerve cells; and a third opinion is that of Marie, who would divide the atrophies into two classes; the first due to neuritis, the second to disease of the anterior horn cells.

Dejerine[31] first published his observations in 1888, a report of five cases, in which changes in the peripheral nerves were found at autopsy. Nonne[32] published the same year a case with atrophy of the hand, with changes in the electrical reactions of the muscles. In this case there was found a degeneration of the intramuscular nerve filaments; the cord showed the lesions of tabes, but was not examined microscopically. In 1889 Dejerine wrote a paper (*loc. cit.*), in which he collected nineteen observations of muscular atrophy with tabes; nine with autopsy. He described changes in the muscle, and particularly in the intramuscular nerve filaments, and considered the changes as due to a chronic neuritis. In the nerve cells he found no changes, but at that time the method of Nissl was not in common use. Many cases have been reported of tabetic amyotrophy, which, examined anatomically, showed neuritis, and of these it will be sufficient to mention Pitres and Vaillard,[33] Oppenheim and Siemerling,[34] Kraus,[35] Jean,[36] and Remak.[37]

Reported cases of muscular atrophy complicating tabes, in which the anterior horn cells were found diseased, are not so frequent. According to Goldscheider,[38] Charcot and Pierret were the first to make this finding in a sixty-year-old tabetic, who for one year before her death presented atrophy of the whole right half of the body. They found the right half of the spinal cord, especially the right anterior horn, distinctly atrophied. Dejerine[39] considered this observation as a case of subacute anterior poliomyelitis, merely coincident with the tabes. Six years later Leyden[40] (1877) published a case of tabetic amyotrophy. The patient was forty-nine years old, and had had tabes for eleven years. For the last five years he had a marked atrophy, beginning in the thenar and hypothenar eminences, and later involving the interossei muscles. The forearms were not much affected, and the biceps and deltoid muscles were normal. The thorax muscles, especially the pectoralis major and serratus anticus major, were very atrophic. The lower extremities were normal. A histological examination of this case showed the typical changes of tabes in the posterior columns, but the nerve cells in the anterior horns, especially in the cervical swelling, were degenerated.

Condoleon reports a case of tabes presenting muscular atrophy, arthropathies, etc., in which he found, besides the lesions of tabes in the posterior columns, a degeneration of the intramuscular nerve filaments and of the anterior horn cells, in segments corresponding to the trophic changes. The nerve trunks and anterior spinal roots were normal. He comes to the conclusion that the degeneration of the anterior horn cells is the primary cause of the atrophy, and explains the fact that the intramuscular nerve filaments are degenerated, by the theory that these being farthest from the cells are first to feel a lessening of the trophic force of the degenerating centre. Dejerine speaks of this as being exceedingly hypothetical. This view of Condoleon, with which Shaffer agrees, that the nucleodistal (Strümpell) portion of a nerve fibre is the first to be affected by disturbances in the cell, has at the present time much to uphold it. In cases of Friedreich's ataxia the degeneration of the fibres in the lateral columns is most marked in the lumbar region which is farthest from their origin, the cells of the cortex. The fibres in the posterior columns are most degenerated in the cervical region, which is farthest from the spinal ganglia, where they originate. Strümpell,[48] in a recent paper, speaks of this in connection with a case of primary lateral sclerosis.

In 1896 Shaffer wrote a paper in which he published cases of amyotrophy in tabes in which, on histological examination, there were found changes in the cells of the anterior horns. Later, in 1898, he published a paper, "Ueber Nervenzellveränderungen des Vorderhorns bei Tabes," in which he agrees with the idea of Charcot and Pierret, that the degeneration of the anterior horn cells is the essential lesion producing the amyotrophies occurring in tabes. He explains the dearth of reported cases showing cell changes by ascribing it to lack of adequate cell-staining methods before the introduction of the Nissl and Golgi stains. As emphasized by Shaffer, the changes in the anterior horns are not those of an anterior poliomyelitis. There are two categories of affections of the anterior horns: one, the ordinary anterior poliomyelitis, which attacks in a rapid and complete manner not only the cells, but the other histological elements of the anterior horns, and which clinically causes rapid wasting, fibrillary contractions, and reactions of degeneration; the other commences with a very fine alteration of the motor, or, as named by Nissl, the stichochrome cell. It is not a process which attacks the cell acutely and extensively, but slowly and progressively. This is seen in some forms of myopathy, such as the toxic, and, if one pleases, the tabetic amyotrophy. Shaffer also points out the clinical features as opposed to anterior poliomyelitis, viz., the slowly progressive character and absence of fibrillary tremor, and of the reaction of degeneration. He concurs in the opinion of Condoleon offered as an explanation of the degeneration of the intramuscular nerves—*i. e.*, the terminal portions of the fibre are the most sensitive to changes in the nutritional power of the cell.

According to Charcot and Pierret this alteration of the gray matter of the anterior horns is consecutive to the lesion in the posterior columns, and constitutes a propagation of this lesion across the posterior cornua to the motor cells. This is explained more in detail by Shaffer[44] as follows: It is well known that the collaterals of the posterior roots have their terminal arborizations about the motor cells of the anterior horns. The vitality of a cell depends on the excitations which reach it, therefore the default of these habitual excitations in tabes through degeneration of the posterior roots produces definite dynamic and histological changes in the anterior horn cells, thus producing the amyotrophies and other trophic changes seen in tabes. In the conclusion of his paper, Shaffer speaks of other conditions which may assist in causing the atrophy, such as disuse and the possible presence in the circulation of syphilitic toxins.

The presence of degenerated cells in the anterior horns of the lumbar region in the first case (W. J. H.) is very important, and strongly confirms the views of those who hold that the amyotrophy in tabes is of cellular origin. The segmental distribution of the most intense atrophy corresponds with the location of the diseased cells in the cord, whereas in other parts of the spinal cord the motor cells are nearly or quite normal. The slowly progressive course of the atrophy is also suggestive of a spinal origin. Another confirmatory point is made by comparing Case I. with Case II.; in the latter there was no atrophy and the anterior horn cells were found to be normal. Unfortunately, the peripheral nerves from the first case (W. J. H.) were not obtained for microscopic study, but the anterior roots seen in sections of the spinal cord stained by the Weigert hæmatoxylin method were found to be normal. This is in consonance with the findings of Shaffer and others, who have reported that the intramuscular nerves were degenerated, but that the nerve trunks and spinal nerve roots were normal. A finding such as this is not uncommon. The peripheral nerves in the portions farthest from the cord may be much degenerated, while the anterior roots are normal. It cannot be denied that the peripheral nerves may have been diseased in Case I. (W. J. H.), but it would be impossible to assert that the peripheral change occurred in advance of that in the nerve cells, and, furthermore, the alteration of the nerve cells of the anterior horns is not like that commonly seen as a reaction at distance, viz., central chromatolysis. As Shaffer says: "The tabetic amyotrophy is not a specific tabetic symptom, though the tabetic process is an etiological factor, but it is rather an associated affection. It is actually a progressive spinal muscular atrophy."

The involvement of the fifth nerve, noted in the first case, is an interesting feature, but not very uncommon in tabes. Spiller[45] has seen a degeneration of the fifth nerve in at least four or five cases of tabes pathologically studied. Most authors refer to the painless dropping out of the teeth in this connection. According to v. Leyden and Goldscheider,[46] the first cases were reported by L'Abbe (1868) and Dolbean (1869). The falling out of the teeth is followed by atrophy of the alveolar processes of both the upper and lower jaws. Necrosis of the jaw may occur and sequestra form. The process is at first painless, but usually infection occurs later. Both the dropping out of the teeth, the necrosis of the jaw, and fracture of the jaw may be early symptoms of tabes, and progress very rapidly. Sensory disturbances in the distribution of the fifth nerve may or may not be present. The most frequent pathological finding is a degeneration of the descending spinal root of the fifth nerve.

I have at present a patient in my private practice who is forty-eight years old, a female, with no history or signs of specific disease. Two years ago she developed symptoms of tabes, lancinating pains, slight ataxia, and partial incontinence of urine. About one year ago she had a purpuric eruption in the same region as the pains, viz., the thighs, and posterior surface of the legs. She now has the diagnostic signs of tabes dorsalis; the lost knee-jerks and lost Achilles jerks, marked ataxia of gait and station, Romberg's symptom, Argyll-Robertson pupils, and delayed sensation in the lower limbs. The most interesting feature of the case is the involvement of the fifth nerve distribution. She has had recurrent attacks of herpes zoster in the region of the distribution of the left supraorbital nerve, with a loss of the supraorbital reflex on the left side. Recently the upper left central incisor tooth became loose and was extracted without pain. Attacks of herpes zoster in the distribution of the spinal nerves have been reported as occurring in tabes, and have been attributed to involvement of the spinal ganglia; but the occurrence of herpes in the distribution of the supraorbital nerve in connection with a loss of the supraorbital reflex and the painless dropping out of the teeth, all pointing to the involvement of the fifth nerve in a tabetic patient, seems to be unique in the literature of the subject.

This patient also has an atrophy of the right half of her tongue, slight but distinct. On protrusion the tongue deviates to the right. The right side of the tongue feels softer than the left, and is more flabby and tooth indented. There is no interference with speech.

The degeneration in the lateral columns in the second case (J. A.) is of importance in connection with the possible exaggeration of the knee-jerks in tabes. The knee-jerks may be preserved in tabes in cases in which the posterior roots in the lumbar region are not affected, or in cases in which the root entrance zone of Westphal is intact, but it has been held that the knee-jerk can never be exaggerated in an uncomplicated case. This view is no doubt correct, but a degeneration of the lateral columns, such as was observed in Case II. (J. A.) might produce no other signs of its presence than slight weakness and exaggeration of the knee-jerks so long as the nerve cells and posterior lumbar roots were capable of transmitting reflex impulses. In such a case we might have the clinical picture of uncomplicated tabes with increased knee-jerks. The development of this sclerosis of the lateral columns might cause the return of the knee-jerks by removing the inhibition of the brain after they have become abolished in the course of the disease, in the same way that a hemiplegia causes the return of the reflex on the paralyzed side. The location and configuration of the areas of sclerosis in the lateral columns show them to be due to interference with the blood supply, which was probably the result of the thickening of the membranes mentioned in the autopsy report. I have not gone farther in my discussion of the involvement of the lateral columns in tabes, as the subject is considered by Dr. A. R. Allen in this number of the BULLETIN.

The presence of swollen axis cylinders in the anterior columns of the spinal cord in the first case (W. J. H.) is a feature worthy of comment. The patient was extremely anæmic, which could probably be accounted for by the chronic infectious process, the psoas abscess. The presence, therefore, of the swollen axis cylinders without any signs of inflammatory change and without any connection directly with the bloodvessels is interesting as showing the earliest change in the spinal cord in anæmia. In most cases the spinal cord changes occur in pernicious anæmia or where anæmia is the chief symptom of the disease, and the alteration is very intense, consisting of many foci of degeneration, these causing secondary degenerations. By a study of Case II. we may conclude that the early alteration of anæmia as shown in the spinal cord consists of a swelling of the axis cylinders.

Pickett[47] recently examined the blood in a number of cases of paresis, and from his observations drew the following conclusion: "When the spinal cord bears the brunt of the disease process in paresis, a simple anæmia with leukocytosis is found; when mental symptoms alone appear, the blood state is not characteristic. Perhaps anæmias or the toxins of anæmias are a factor of the cord lesions even of paresis." The connection between chronic degenerative conditions of the central nervous system and degenerative blood conditions is well worthy of further observation.

For the pathological material and many valuable suggestions in the preparation of this paper, I am indebted to Dr. William G. Spiller.

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A PATHOLOGICAL STUDY OF ACUTE MYELITIS.[1]

With a Report of Two Cases.

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of Philadelphia.)

Notwithstanding the fact that the subject of acute myelitis has been extensively studied, and that a large amount of experimental work has been done with a view of throwing some light upon the nature of the process, much doubt surrounds the question, and a number of different opinions are still held as to the exact character of the morbid process which gives rise to the symptoms commonly known as acute myelitis. Marie[23] has gone so far as to say that primary myelitis does not exist, and he with Schiff[35] viewed the process as partly infectious and partly the result of an intoxication. It is looked upon as an inflammatory process by Strümpell,[39] Obersteiner,[28] Bielschowsky,[48] and others. On the other hand, Tietzen[40] advanced the theory that the myelitic foci have a mechanical origin, that is to say, that they are the result of the cutting off of the blood supply. Mager,[64] who has made an exhaustive study of the subject, having collected a large number of cases, believes that disease of the blood-vessels is essential to the anatomical diagnosis of myelitis, and that inflammatory change of the spinal cord is present only when changes in the vessels are demonstrated. Leyden and Goldscheider[19] recognize three stages in the process present in acute myelitis: (1) general softening; (2) yellow softening; (3) resorption. In Oppenheim's[29] opinion acute myelitis is a diffuse inflammatory and softening process. Obersteiner and Redlich[30] have stated as their opinion that in some cases the change in the spinal cord is confined to the nerve elements, the connective tissue remaining intact, a condition which they call parenchymatous inflammation, or there may be acute degeneration associated with an inflammatory process. Brissaud[49] has described an apoplectic myelitis. Schmaus[33] divides the process into (1) a condition of swelling in which there is œdema which may or may not depend upon inflammation; (2) parenchymatous degeneration in which the nerve elements are destroyed without involvement of the neuroglia; (3) an infiltration process in which there is change in the vessels without softening; (4) spinal softening, which may be primarily inflammatory, or secondary from inflammation; (5) suppurative inflammation of the spinal cord. Leyden and Redlich[21] differentiate two groups of acute myelitis: (1) pure transverse myelitis; (2) disseminated myelitis, and conclude that in those cases in which thrombosis or embolism can be excluded, an inflammatory process is the probable cause of the myelitis.

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[1] Read before the Section on General Medicine of the College of Physicians of Philadelphia, January 9, 1905.

Grasset and Rauzier[9] believe that in the first stage of acute myelitis there are intense congestion, escape of leukocytes, infiltration into the gray substance, and swelling of the nerve elements; in the second stage proliferation of the connective tissue, and frequently formation of fatty granules, and finally, softening.

B. Kahlden[18] states that there may be degeneration without disease of the bloodvessels in the cord, as in other organs.

It will thus be seen that there exists a diversity of opinion on this subject among well-known authorities, and it will, therefore, not be out of place to cite the following two cases which throw some important light upon the question.

Case I.—L. R., a married woman, aged twenty-nine years, was admitted to the wards of St. Agnes' Hospital, in the service of Dr. Aloysius O. J. Kelly, December 31, 1903. A week previous to her admission she began to suffer from burning pains in the epigastrium, which extended to the back and which were associated with vomiting. On the following day she complained of pain in both legs, extending from the tips of the toes up on the trunk to a level about half an inch above the umbilicus. Over the same distribution she experienced a sensation of pins and needles. On the same day weakness of the legs developed, and in a few hours there was complete motor paralysis.

When examined on admission, six days after the onset of the paralysis, she was unable to move her legs at all. There was no paralysis of the arms. The urine was retained, and the feces were passed involuntarily.

The knee-jerks could not be elicited, and there was no clonus, no ankle-jerk, and no Babinski phenomenon. The plantar reflex, however, was present on both sides.

There was distinct alteration in sensation to pain in the legs, but on account of the poor mental condition of the patient the exact distribution of the loss of sensation and its exact character could not be ascertained.

Kernig's sign was not present, and there was no tenderness of the spine. When the patient was moved she complained of pain generally, but especially across the upper part of the chest. There was on the inner side of the right foot a bleb one and three-quarters by one inch in diameter, evidently trophic in origin.

After admission the patient vomited almost incessantly for several days. There was a cough, and the mucus which collected in her throat was expectorated with difficulty. However, the lungs and heart showed no abnormalities.

On the tenth day of the disease there was marked abdominal distention, which was partly relieved by the rectal tube. At this time the patient complained of itching and burning pain in both arms, extending to the finger-tips.

On the next day, the eleventh of the disease, the vomiting stopped, and she complained of pain in the head, ears, arms, and chest. Her condition grew from bad to worse, and on January 22, 1904, twenty days after the onset of the disease, she died.

On admission the temperature was normal, pulse 122, respirations 28. The disease ran an irregular febrile course, the temperature never rising above 102° until the last day, when it reached 106°. An examination of the blood, made by Dr. J. J. Kane, gave the following results: red blood cells, 4,960,000; white blood cells, 4240. The Widal reaction was negative, and the examination of the urine at no time showed any deviation from the normal.

The family history was negative. There was no history of phthisis, cancer, or syphilis. She began menstruating at fifteen, and was always regular, the periods lasting two or three days. There was great pain on the day preceding, and during the first day of the flow. On admission she was in the fifth month of pregnancy. The child was viable, and remained so until the time of the death of the mother.

Only the spinal cord of this case was obtained. It was hardened in Müller's fluid, and sections were taken from various levels of the cord and stained by the Weigert and Marchi methods and by Ehrlich's hæmatoxylin. There was an intense round-cell infiltration of the pia throughout the whole length of the cord, and also in many places within the cord. The process was more marked in the thoracic and lumbar regions. At about the level of the second lumbar segment there was an area of softening extending about three-quarters of an inch. Scattered throughout the entire cord as far as the midcervical region—higher than which the cord was not obtained—there existed numerous foci of inflammation within the cord.

In the cervical enlargement there was in the direct pyramidal tract on one side a focus which was composed of numerous bloodvessels surrounded by a round-cell infiltration, and some of these vessels were obliterated. The walls of the vessels were somewhat thickened. In this focus the nerve elements were entirely destroyed. In the posterior columns at this level there was a second focus on the opposite side involving a large part of the posterior columns. This focus was histologically identical with the one situated anteriorly.

In the upper thoracic region in the crossed pyramidal tract on one side there was a small, comparatively recent hemorrhage around which was a moderate number of round cells. The process was more severe in the posterior columns at this level, and in one horn the process was especially marked around a number of bloodvessels, some of which were obliterated.

In the middle thoracic region the process was more intense and involved pretty generally the whole cross-section of the cord, but was especially marked in the anterior and posterior horns on one side. There was a hemorrhage in the posterior portion of the anterior horn on one side at this level.

In the lower thoracic region the process was still more extensive, and here scarcely any part of the cord was exempt.

In the lumbar region the pia was much thickened posteriorly, and the softening had destroyed a large part of the posterior half of the cord for an extent of about three-quarters of an inch.

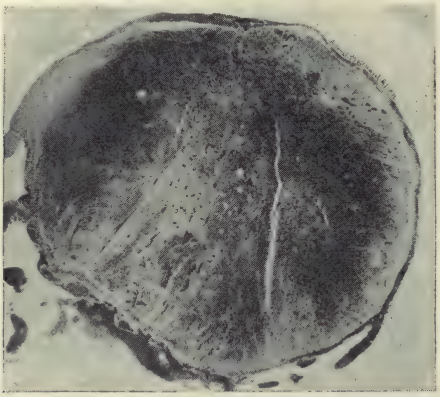

Fig. 1.—Section from the thoracic region of the spinal cord stained by the Weigert hæmatoxylin method. The differentiation between white and gray matter has disappeared, because of intense myelitis. The light areas represent complete degeneration.

The foci of inflammation failed to take the Weigert stain, as shown in the accompanying photograph. (Fig. 1.) The border-line between the healthy and diseased areas was more or less sharply defined. In the parts where the process was less intense some nerve tissue was retained.

Sections from the thoracic and cervical regions studied by the Marchi method showed an intense and recent diffuse degeneration. Numerous granular cells were seen in the areas where the disease was most marked. (Fig. 2.) In the other parts of the cord at these levels the myelin was diffusely degenerated. The Nissl method could not be employed, but the nerve cells in the anterior horns of the cervical region, as stained by acid fuchsin, showed much alteration. In some nerve cells the nucleus was displaced far to the periphery of the cell, and some of the cells were much swollen.

The cells, which were present in the foci of inflammation, were mononuclear. There was, however, present a number of cells somewhat larger, the protoplasm of which appeared to be granular. These were mingled with the round cells, and were looked upon as being *corps granuleux* or fatty granular cells (Fettkörnchenzellen).

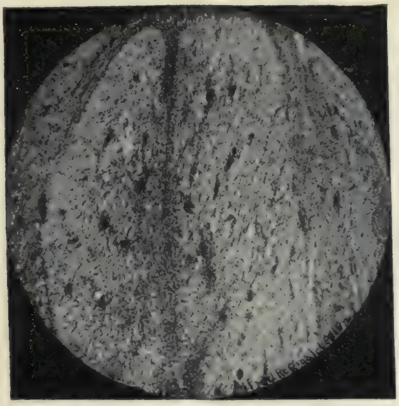

Fig. 2.—Section from the thoracic region, showing intense round-cell infiltration within the cord. The bloodvessels are unusually numerous.

Case II.—The second case is that of a single woman, aged thirty-nine years, a shirtmaker by occupation. She was admitted to the wards of St. Agnes' Hospital on November 13, 1903, in the service of Dr. Aloysius O. J. Kelly. Her family history was as follows: The mother died during the menopause of an unknown cause. The father was living at the age of seventy-five, and was asthmatic. All of her brothers and sisters suffered from phthisis. There was no history of cancer or syphilis. She had had measles, diphtheria, scarlet fever, and acute articular rheumatism when young. For six years previous to her twelfth year she had suffered from quinsy each winter. In 1901 she had a severe attack of influenza. The menstrual periods began at ten years, and were always irregular, sometimes bimonthly, of three or five days' duration, and abundant. She had suffered from leucorrhœa for the past four years. Twelve years ago she had typhoid fever, and a second attack four years ago. She was delirious for six weeks during the first attack, and for some time afterward her mind was not in good condition; and in the last attack she was delirious for seven weeks, and her mind was feeble for six months afterward.

On October 23d, twenty days before her admission to the hospital, she had what she considered an acute attack of rheumatism, which prevented her from going to her work. On account of the mental condition of the patient the history of the onset and progress of her illness however, could not be obtained.

Upon admission examination showed a well-developed woman, aged thirty-nine years, fair skinned and rosy-cheeked. The examination of the lungs and heart was negative. The liver was slightly enlarged, extending about one-half a finger's breadth below the margin of the ribs. The spleen was normal. Palpation of the abdomen apparently caused pain.

When examined by myself six days later she was paraplegic, but was able to move her feet slightly and to turn them in or out. The movements, however, were feeble, and there was no power above the knees.

There was total anæsthesia below the sixth rib in front, and from a level four inches lower in the back. There was incontinence of urine and defecation was involuntary. The pupils were equal, and reacted normally, and the tongue was protruded straight. She moved her head freely and easily, although any motion of the head appeared to give her pain.

The knee-jerks were absent, there were no abdominal reflexes, but Babinski's phenomenon could be developed. At this time there was no involvement of the arms. Three days later, however, the extensor and flexor muscles of the arms and hands became partially paralyzed, and in three days the paralysis of the upper limbs became total.

Dr. T. W. Tally states that his examination of abdominal organs was negative. The blood count showed 4,360,000 red cells, 8720 white cells, and hæmoglobin 80 per ecnt. The course of the disease was practically afebrile. The examination of the urine was negative.

On November 25th, thirty-two days after the onset of her symptoms, and four days after the extension of the palsy to the arms, the patient became unconscious and died.

Only the spinal cord was obtained for study. This was placed in Müller's fluid and sections taken from various levels and stained by the Weigert and Marchi methods and by carmine and acid fuchsin. A study of the numerous sections failed to reveal the presence of any round-cell infiltration either in the pia or in the cord itself. Unfortunately the Nissl method could not be employed, but the nerve cells in the anterior horns of the cervical region, as stained by acid fuchsin, appeared to be normal.

In the thoracic region, however, the nerve cells of the anterior horns were much swollen. Otherwise the examination of the cord at this level was negative.

In the lumbar region the cells were much altered, the nuclei being displaced and the cells themselves swollen.

No round-cell infiltration of the cord or pia could be seen.

By the Marchi method in the lower thoracic region the myelin sheaths were pretty generally stained everywhere. There was, however, an absence of the granulations and of the compound granular cells, which are characteristic of acute degeneration, as shown by the Marchi method. It should be stated, however, that the cord had been hardened in Müller's fluid for several months, and had remained in osmic acid for at least seven or eight days. This throws some doubt upon the value of these findings, and the staining found may, in view of these facts, be merely an artefact, especially since subsequent study of sections placed in osmic acid solution for five days failed to disclose the presence of any degeneration.

The first case presented the usual pathological change of acute myelitis—*i. e.*, the presence of areas of softening, change in the bloodvessel walls, round-cell infiltration, and destruction of the nerve elements. There was found, however, as studied by the Marchi method, a general acute degeneration of the nerve elements which was not confined to the foci of softening, nor to the cord in the neighborhood of the diseased bloodvessels. This is important and interesting in relation to the study of the character of the pathological process of acute myelitis as relating to the problem whether true acute myelitis depends always upon an inflammatory process, the mechanical results of disease of the bloodvessels, a thrombotic or embolic process, or a degenerative process. While there is no doubt that there was present in this case an inflammatory lesion, I think it cannot be concluded that this was primary, and that the change in the nerve tissue was caused by the bloodvessel change alone. If the disease of the bloodvessels was primary in this case the diseased nerve elements would be more or less confined to the neighborhood of the diseased vessels, and a different explanation would be required to account for the presence of a marked and widespread degeneration of that portion of the cord which was far removed from the diseased bloodvessels. It is suggestive, at least in this case, that the cord and bloodvessels alike were attacked by a toxin or by some infectious agent which brought about an inflammatory process in the bloodvessels, and at the same time a degenerative process in the nerve elements. Marie,[23] Schiff,[35] and Küstermann[16] have already advanced this explanation of myelitis.

In the second case reported in this paper the findings are, to say the least, unique. In a woman who suffered from motor and sensory paralysis below the waist and probably of acute onset, with death in six weeks, a study of the cord showed an entire absence of inflammatory change beyond the presence of a few recent hemorrhages in the posterior horns at certain levels, which could readily have occurred during the agony of death; the presence of a decided change in the cells of the anterior horns below the cervical region, and a moderate degree of degeneration in the sciatic nerve. In this case by the method of Marchi there existed possibly a widespread degenerative process, which had not progressed sufficiently to break up the axis cylinders and to produce the granular cells characteristic of acute degeneration. If it is true that the staining of medullary sheaths by the Marchi method in this case is an evidence of beginning degeneration, then it may be looked upon as an important finding, and as an indication, if not a proof, that acute degenerative change may occur independently of a circulatory or inflammatory disturbance. If it is a mistake to look upon this staining of the myelin as evidence of acute degeneration, then it is difficult to make the clinical phenomena and the pathological findings agree. The symptoms were undoubtedly those commonly ascribed to acute myelitis, that is, paraplegia, loss of sensation, involvement of the sphincters, and trophic disturbances. Excluding the possible degeneration of the myelin as a pathological basis for these symptoms, there was nothing abnormal found beyond the degeneration in the nerve cells and the sciatic nerve, and this is entirely insufficient to explain the clinical phenomena. I may in this connection allude to Landry's paralysis, in which lesions explanatory of the paralysis are sometimes absent.

I have studied fifty-two cases of so-called acute myelitis, with autopsy, in the literature, which, with the two cases above reported, make fifty-four cases with autopsy in which the clinical symptoms were very similar. I have excluded the cases of abscess of the cord, of syphilitic disease of the cord, and the apoplectic cases.

These cases can be divided rather readily into two distinct classes. In the first class there is evidence of an inflammatory process in which the bloodvessels are involved, in which there is infiltration of the tissue, and more or less destruction of the nerve elements. In the second class the appearances of inflammation are absent and the degeneration of the nerve elements seems to be entirely independent of it. These latter cases are greatly in the minority.

Buchholz[6] describes a case in which the axis cylinders were destroyed, and in which amyloid bodies were present. He looks upon these amyloid bodies as the products of the degeneration of the nerve fibres. In this case inflammatory infiltration played a very insignificant role.

In a case described by Lauenstein,[20] in which the patient died in seventy-two hours, there was swelling of the axis cylinders, and homogeneous masses which did not take the stain were found in the neighborhood of the vessels without having any communication with the perivascular spaces. The bloodvessel walls were in no respect changed, and the neuroglia showed no increase in the nuclei.

In Küstermann's[16] case, a woman, aged forty-eight years, who died in nineteen weeks, the connective tissue of the cord was entirely destroyed, the nerve fibres were absent, fat drops and granular cells were present, but there was no material change in the vessels or the vessel walls except in the pia, the walls of which were thickened and in some places obliterated. Küstermann looks upon this case as of infectious origin, and expressed the opinion that in some cases the vessels become infected and are then the origin of the disease, while in other cases the infectious irritant passes through the vessel walls without altering them and brings about disease of the substance of the cord itself. In Verger and Abadie's[43] case the process appeared to be a degenerative one. There was in this case also a peripheral neuritis. Walter K. Hunter[11] cites a case in a man, aged forty-five years, who died two days after the development of the paralysis. At the post-mortem examination there was an extensive degree of softening which the author concludes was post-mortem. There was no evidence of an inflammatory process anywhere, but the change observed was in the nature of a degenerative softening. Fraenkel's diplococcus was found in the softened areas. These comprise the cases found in the literature in which with the clinical manifestations of acute myelitis there was found post-mortem only a degenerative process.

Real progress has been made in this subject by experimental work. Hoche[13] experimented on animals with toxic and non-toxic substances, and was unable to decide whether the change induced was a mechanical effect of a foreign body or a result of the action of a chemical poison. He was able to demonstrate the presence of bacteria in the diseased portions of the spinal cord, and looked upon this as evidence against the theory that localized spinal disease was the result of a poison circulating generally in the blood. In his experimental cases the diseased foci were found only where the micro-organisms were present. He produced by combining embolism with injections of pathogenic micro-organisms, an ischæmic change as well as a condition which could be designated as acute infectious myelitis.

Marinesco[27] experimented with the staphylococcus, streptococcus, pneumococcus, meningococcus, influenza bacillus, bacillus of typhoid fever, the colon bacillus, the tubercle bacillus, and the leprous bacillus. He concluded that there are three classes of infection: (1) infections which, properly speaking, are not myelitis, but in which the microbes accumulating in the lymph spaces and in the arachnoid cavity attack the nervous tissue, probably by secreting toxic substances without determining a vascular or leukocytic infection; (2) infectious meningomyelitis caused by microbes injected into the arachnoid canal;

(3) conditions resembling poliomyelitis caused by injecting the influenza bacillus and the streptococcus. The lesions found consisted of nodular inflammation, swelling of the neuroglia, increase in the nuclei, and chromatolysis and achromatolysis of the nerve cells. Somewhat similar experiments have been performed by Roger,[32] who produced degeneration of the bloodvessels and hemorrhagic foci; by Babinski and Charrin,[2] who produced motor paralysis in rabbits without pathological findings; by Bourges,[3] who demonstrated experimentally alterations in the gray substance and the presence of granular bodies and vascular lesions, and by Homen,[24] Zappert,[44] Friedman,[7] Hochhaus,[15] Henriquez and Helion,[14] Ballet and Lebon,[1] Bouchard,[4] Moret and Rispal,[25] Widal and Bezançon,[41] and Vincent.[42] Maget[26] produced medullary lesions by inoculating animals with cancer cells, and Sabrezes and Mongour[37] by injecting blood from a case of infectious icterus produced a diffuse myelitic change in the anterior horns in animals. The histological picture found in experimental myelitis agrees with that found in the majority of cases of acute myelitis in man.

Infection seems to be an important element in the causation of this disease. In a case reported by Lépine,[61] which occurred during typhoid fever, there was found an intense proliferation of the neuroglia with the classical changes of acute myelitis.

Marinesco,[65] who believes that acute myelitis is an infectious condition, reported six cases, in two of which the streptococcus was found; in one the pneumococcus, and in one a bacillus resembling the anthrax bacillus. Auche and Hobbs,[45] in a case of myelitis of seventeen days' duration, developing during an attack of variola, found diplococci and micrococci in a fibropurulent exudate around the cord. There was further evidence of acute inflammation, such as infiltration of the perivascular spaces, bloodvessel change, and alterations in the neuroglia and nerve fibres.

Achard and Guinon[46] describe a case which they believe was of an infectious character. Histologically there were large numbers of cells, probably granular bodies, infiltrating the nerve elements, especially around the bloodvessels. There was also in this case an optic neuritis.

Marinesco and Oettinger[62] describe a case occurring in a man, aged twenty years, who was suffering from variola and who died four days after the development of the paralysis. In this case the streptococcus was found in the ependymal canal, and vascular lesions, alteration in the nerve cells, hemorrhages, and change in the axis cylinders were present. They looked upon this case as one of acute ascending paralysis or Landry's disease, and concluded that this affection is, in the majority of cases, due to a microbic agent acting upon the nervous system. Stuart[70] found the staphylococcus in the spinal cord of a man, aged eighteen years, who died in three weeks after the development of a flaccid paraplegia. There were present perivascular infiltration of mononuclear leukocytes.

Barlow[50] described a case of acute myelitis occurring in a man, aged twenty-three years, who was suffering from measles, and which came to a fatal termination eleven days after the development of the paralysis. There was marked vascular change and leukocytic infiltration. The nerve cells, nerve fibres, and neuroglia were unaffected.

In a patient, aged nineteen years, reported by Schiff,[35] there suddenly developed in the ninth day of typhoid fever general weakness, paralysis of the sphincters, loss of motion in the legs, diminution of the reflexes, and irregular pupillary reaction.

Anæsthesia in all the extremities and the thorax developed later, and still later the arms became weak. Death occurred at the end of eighteen hours, and the autopsy revealed the presence of multiple hemorrhages throughout the spinal cord. The bloodvessels were dilated and filled with blood; there was marked degeneration of the ganglion cells in the anterior horns of the cervical region, the axis cylinders were swollen, and there was an overgrowth in the pia. Micro-organisms were not found.

A case is reported by Fürstner[8] in which a girl, aged sixteen years, developed acute myelitis which proved to be fatal in twenty-four days. The autopsy showed the presence of granular cells, degenerated myelin, but no round-cell infiltration. The case was not looked upon as an inflammatory one. The pneumococcus was present.

The experimental cases failed to show the presence of organisms in the spinal cord after the ninth or tenth day of the disease. In animals who died before this time the micro-organisms were frequently found in the cord substance. This fact probably accounts for the failure to find an infectious organism in more cases of acute myelitis.

In my first case the myelitis developed during the fourth month of pregnancy. While myelitis may occur in pregnancy, it certainly is not a frequent occurrence, as few such cases have been met with in a study of the literature. Leyden, however, considered the association of pregnancy and myelitis of sufficient importance to mention the former as an etiological factor. Schnitzer[36] cited a case of myelitis developing in the sixth month of pregnancy in a woman, aged thirty-three years, who died in two months after the onset of the paralysis. At the post-mortem examination the dura was thickened and the spinal cord narrowed in the mid-dorsal region, and there was a white, gelatinous mass surrounding the cord.

Kreutzman[17] describes a case of paralysis occurring in a woman, aged twenty-four years, during her first pregnancy, but the course of the case was not given further than to state that the child was born.

In a case reported by Alexander Bruce[5] a pregnant woman, aged thirty-one years, during an attack of vomiting suddenly fell to the floor complaining of intense pain all over the body. Power was lost in both legs and in her right arm. There was anæsthesia, paralysis of the bladder and rectum, and the patient died in two months. The cord was softened in the thoracic region. A tubular hemorrhage was found throughout the length of the cord from the sixth cervical to the first lumbar segments. This tube was probably a malformation of the central canal, and when the hemorrhage occurred it burst through the wall of the tube.

Collins[52] cites a case of a woman, aged twenty years, who, during her first pregnancy, quite rapidly lost power in her legs and developed bed-sores. The knee-jerks were absent, the legs spastic, and the sphincters were paralyzed. There was no sensory change noted. The woman died at the expiration of three months. In the cord there were found foci of necrosis around which the blood-vessels were thickened, the connective tissue proliferated, and there was round-cell infiltration in the perivascular spaces. The typhoid bacilli were found. By the Nissl method the cells in the anterior horns were degenerated.

Bruce cites Oliver's[31] case, that of a woman, aged twenty years, who in her second pregnancy became paraplegic. Labor went on without difficulty or pain, and the patient died in ten days. The autopsy showed compression of the spinal cord.

Other cases have been reported by Imbert-Gombeyre, D'Arcy, and Trevalyan, but the references were not obtainable.

It may be concluded, I believe, that (1) the most frequent pathological findings in acute myelitis consists of necrotic foci, thickened bloodvessels, round-cell infiltration, small hemorrhages, destruction of the nerve fibres in the neighborhood of the diseased bloodvessels, alteration of the nerve cells, and the presence of granular cells; (2) more rarely there may be degeneration of the nerve elements without evidences of inflammation; (3) there may be cases in which these two processes seem to be associated; (4) that infection plays an important role in the production of acute myelitis; (5) that the cause of acute myelitis may be either a micro-organism, the products of a micro-organism, or a toxin.

Cases in which inflammation of the cord was present:

Auche and Hobbs[45] (*Compte rend. de la Societe Biolog.*, 1894, No. 1, p. 703).—Duration, seventeen days; perivascular infiltration, slight vascular change; alteration of the nerve elements; diplococcus found; variola.

Achard and Guinon[46] (*Archiv. de med. Experimentale,* 1889, p. 696).—Duration, six months; granular bodies; vascular infiltration; secondary degeneration; optic neuritis.

Bainit[47] (*Deut. Zeit. f. Nervenheilk.*, 1901, No. 19).—Softening; degenerated vessels; secondary degeneration; nerve elements destroyed.

Bielschowsky[48] [38] (two cases) (*Neurologisches Centralbl.*, 1899, No. 18, p. 1088).—1. Duration, three weeks; hemorrhages; vascular change; degeneration of nerve tissue; leukocytic infiltration. 2. Duration, two months; inflammatory process; involvement of nerve tissue.

Brissaud[49] (*Gaz. hebdom. de Med.*, February, 1902, p. 133).—Necrosis; "lükenfeld;" hypertrophy of nerve fibres attributed to vascular lesions; apoplectic softening.

Barlow[50] (*Med. and Chirur. Transactions*, London, 1887, No. 72, p. 77).—Duration, eleven days; vascular change; softening; no degeneration of nerve tissue, cells, or neuroglia; leukocytic infiltration.

Buzzard and Russell[51] (*Medical Press and Circular*, London, 1898, p. 329).—Duration, forty-two days; leukocytic infiltration (diplococcus in meninges); engorgement of vessels; cellular infiltration; hemorrhages.

Collins[52] (*Journal of Nervous and Mental Disease*, 1902, p. 705).—Duration, three months; necrotic area; vascular change; proliferation of connective tissue; round-cell infiltration; tubercle bacilli found at the margin of necrotic area; nerve tissue involved.

Dreschfeld[53] (*Lancet*, 1882, vol. i. p. 82).—Duration, six weeks; engorged vessels; granular and round cells; secondary degeneration.

Evans[54] (*Lancet*, 1894, No. 2, p. 683).—Duration, four days; leukocytes in anterior horns; dilated vessels; disappearance of nerve fibres.

Eisenlohr[55] (Virchow's *Archiv*, 1878, p. 82).—Duration, eight weeks; neuroglia thickened; axis cylinders degenerated; vascular change.

Fragotte[56] (*Archiv f. Neurologie*, 1890, p. 46).—Degeneration of the nerve fibres; sclerosed vessels; hemorrhages; Deiters' cells.

Hochhaus[57] (*Deut. Zeit. f. Nervenheilk.*, 1899, four cases).—1. Duration, thirty days; diseased vessels; epithelioid cells; destroyed nerve vessels. 2. Duration, some weeks; inflammatory foci; round-cell infiltration; granular detritus. 3. Duration, twenty-six days; inflammatory foci; round-cell

infiltration; white matter destroyed. 4. Duration, two months and twenty-five days; overgrowth of neuroglia; round-cell infiltration; nerve tissue destroyed; in these places the vascular disease was not confined to the myelitic foci.

Henneberg[58] (*Centralblatt f. Nervenheilk. und Psychiatrie,* 1899, vol. x. p. 8).—Duration, eight weeks; inflammatory process (specific).

Jacobeus[59] (*Zeit. f. klin. Med.*, 1898, No. 36, p. 355).—Duration, eight days; destruction of white substance; dilated vessels; hemorrhages.

Knapp[60] (*Neurologisches Centralblatt,* 1885, p. 502).—Disease of vessels; softening; round-cell infiltration.

Lépine[61] (*Lyon Med.*, 1903, p. 742).—Acute myelitis, with intense proliferation of neuroglia; variola.

Marinesco and Ottinger[62] (*La Semaine Med.*, 1895, p. 45).—Duration, forty-six days; vascular lesions; polynuclear and mononuclear leukocytic infiltration; leukocytes filled with the diplococcus in the anterior horns; prolongations of the axis cylinders ruptured; perivascular infiltration; small foci and vascular disease in medulla.

Mouratoff[63] (*Revue de Med.*, January, 1903, p. 40, two cases).—1. Duration, eight days; acute degeneration (Marchi), vascular change, *i. e.*, destruction and perivascular exudation; parenchymatous neuritis. 2. Duration, two days; hyperæmia and perivascular infiltration; axis cylinders swollen; fat drops.

Mager[64] (*Arbeiten aus dem Neurol. Institute,* 1900, seven cases).—1. Duration, nine days; vascular disease; infiltration of adventitia. 2. Duration, twenty-eight days; necrotic foci; vascular disease. 3. Duration, forty-two days; necrotic foci; vascular changes; infiltration. 4. Duration, twenty-two days; necrotic foci; vascular disease;

infiltration of adventitia. 5. Duration, twenty days; necrotic foci; infiltration; vascular disease. 6. Duration, thirty-two days; disseminated foci; granular cells; degenerated vessels. 7. Duration, six months; infiltration of adventitia; thickened vessels; foci of softening.

Marinesco[65] (*Neurologisches Centralblatt,* 1901, p. 282, six cases).—In four cases micro-organisms were found.

Moeli[66] (*Archiv f. Psychiatrie,* 1880, p. 757).—Duration, fourteen months; vessel changes; cells in anterior horns changed; nerve fibres destroyed; ascending and descending degeneration.

Nageotte[67] (*Nouvelle Iconogr. de la Salpetriere,* 1895, vol. viii. p. 221, two cases).—1. Duration, six weeks; vascular disease; diapedesis of leukocytes; medullary lesions. 2. Duration, five weeks; round-cell infiltration; myelitic foci in relation to vessels; softening; destroyed axis cylinders; gray matter involved.

Pfeiffer[68] (*Deut. med. Woch.*, 1896, No. 22, p. 41, two cases).—1. Duration, six weeks; vascular change; destruction of nerve elements. 2. Scattered foci; vascular change.

Peabody[69] (*Medical Record,* 1883, p. 122, two cases).—1. Duration, seven days; vascular infiltration and thickening; cells in anterior horns degenerated; white matter normal. 2. Duration, four days; inflammatory change in posterior horns; nerve tissue degenerated.

Stuart[70] (*Revue de Neurolog. et Psychiatrie,* 1903, No. 6).—Duration, three weeks; foci in gray and white matter; perivascular infiltration of mononuclear leukocytes; staphylococci in abundance.

Singer[71] (*Brain,* 1902, p. 332, two cases).—1. Duration, seven or eight weeks; softening; ascending and descending degeneration; vascular change; cellular infiltration; no neuroglia change;

caused by deficient blood supply. 2. Duration, seven weeks; cellular infiltration; proliferation of the perivascular connective tissue; no leukocytic infiltration; caused by deficient blood supply.

Strümpell[72] (*Archiv f. Psychiatrie,* 1899, No. 31, p. 848).—Duration, two and one-half months; softening; fatty granular cells; accumulations of myelin; usual changes of acute myelitis; with ascending and descending degeneration; staphylococcus present.

Tschistowitsch[73] (*Pathologisch. Anat.; Arbeiten, Orth.,* 1903, p. 32).—Duration, fifty-seven days; softening, above and below which the axis cylinders were swollen and destroyed; round-cell infiltration, especially around the vessels; caseous tubercles.

Taurelli and Salimbeni[74] (*Revue Neurologique,* 1895, p. 212, three cases).—Small-cell infiltration.

Webber[75] (*Journal of Nervous and Mental Disease,* Chicago, new series, 1880, No. 5, p. 669).—Duration, thirteen days; vascular change and parenchymatous degeneration; small cells; granular contents.

Williamson[76] (*Lancet,* 1895, vol. ii.).—Duration, four months; round-cell infiltration; granular cells; leukocytes; secondary degeneration.

Wersilow[77] (*Neurologisches Centralbl.,* 1898, No. 17, p. 191).—Duration, eight months; infiltration of the vessels; inflammatory process.

Weill and Gallavardin[78] (*Revue Neurologique,* 1903, p. 999).—Epithelioid cells around vessels and infiltrating cord.

I am indebted to Dr. W. G. Spiller for much assistance in the study of the sections.

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COMBINED PSEUDOSYSTEMIC DISEASE, WITH SPECIAL REFERENCE TO ANNULAR DEGENERATION.[1]

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General Hospital.)

The following study of pseudosystemic sclerosis deals especially with that rare form presenting an unbroken ring of degeneration around the periphery of the spinal cord. It is not infrequent to find in sections stained by the Marchi and other methods a dark annular peripheral zone of varying width. This is usually an artefact, and not a true degeneration. It can be caused by allowing the section to remain in the hardening fluid too long, thus permitting the peripheral portion of the cord to become more penetrated by the fluid than the central portion, and it is also noted in sections where the osmic acid has not penetrated well toward the centre.

In cases of recent degeneration implicating the entire transverse area of the cord, sections stained by the Marchi method may show the characteristic reaction only at the periphery, if the osmic acid has not penetrated to the centre.

In discussing a true degeneration of this annular variety it is necessary to revert to the blood supply of the part. The arteries of the cord have most conveniently been divided into two classes, centrifugal and centripetal. The centrifugal system has its origin in the arteria sulci, from the anterior spinal artery, some 200 to 300 in number, which leave the main trunk at right angles, penetrating the anterior median sulcus as far as the commissure, where they turn sharply to the right or left (rarely if ever bifurcating—Kadyi). In this location they are known as the commissural arteries. Each commissural artery gives off the following branches:

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[1] Read before the Philadelphia Neurological Society, January 27, 1904.

(*a*) Anastomotic twigs, one ascending directly and uniting with a descending branch of the corresponding commissural artery, one descending to unite with the ascending twig below. By corresponding commissural arteries I mean arteries which have turned to the same side of the cord. These anastomoses form, on each side and anteriorly to the central canal, a longitudinal artery considered in former times a vein.

(*b*) The anterior cornual branch, which gives a rich blood supply to the anterior horn, penetrating for a short distance into the white substance surrounding. This is the most considerable and important of the branches.

(*c*) A slender branch, passing slightly outward and backward to the Stilling-Clarke vesicular column. Obersteiner[1] speaks of this branch as being the exclusive supply to the vesicular column, while Schäfer[2] makes "special mention" of a branch artery entering the posterior median fissure and supplying the vesicular column.

So much for the centrifugal blood supply.

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[1] Obersteiner. Anleitung beim Studium des Baues der Nervösen Centralorgane im gesunden und kranken Zustande.

[2] E. A. Schäfer. The Spinal Cord and Brain. (Quain's Anatomy.)

The centripetal supply comes from the posterior spinal arteries which form, by division and irregular anastomoses, as well as by the addition of arteries entering the spinal canal with the anterior and posterior roots, a rich arterial investment around the whole circumference. This blood supply is greatest in the posterior columns and of small amount in the anterior aspect of the cord, which is chiefly supplied by the anterior radicular arteries and the anterior cornual branch of the commissural artery. The centripetal supply, which has aptly been termed the arteria corona, sends its branches into the substance of the cord in direction coincident with its radii. As observed above, the branches to the posterior columns are most considerable, and are usually five in number, one penetrating the pial insinuation in the posterior median line, two others on either side of this. The posterior column arteries are not only of larger calibre than those on the lateral aspect of the cord, but they also penetrate to a much greater distance, for the reason, I presume, that the arteries of the gray substance do not project into the posterior columns as they do into the lateral and anterolateral columns, although some authorities hold that the most anterior part of the posterior columns is supplied by twigs leaving the adjacent gray substance. These authorities give this as a reason why the cornu-commissural tract remains so often undegenerated in posterior system disease of vascular origin. There are three fairly constant arteries passing into the white substance of the lateral columns, and in each posterior root is an artery which supplies the posterior third of the gray matter. It seems to me impossible to give any diagrammatic plan of the posterior system of arteries, although such a plan has been given in a number of text-books. It only requires a cursory glance at the posterior aspect of the spinal cord to see how utterly irregular and chaotic the arterial supply is.

The case I am reporting is from Dr. Potts' service in the Philadelphia General Hospital, and I am much indebted to him for the pathological material and clinical notes. The history is as follows:

Mary L., aged twenty-two years, white, born in Germany; admitted May 28, 1902. Father dead; cause unknown. Mother died of "dropsy." Two brothers and three sisters living. Two years ago suffered from "sick stomach." Vomited a great deal. Her present complaint on admission to hospital is that she cannot walk because of "stiff legs," and is "sick at the stomach."

History of Present Illness. About a year ago was taken sick and could not work. This inability continued for ten weeks, when she was brought to the hospital unable to walk.

Physical Examination. Patient fairly well nourished; is quite nervous. Pupils equal and respond to light. Legs wasted. Reflexes exaggerated and ankle-clonus present. Six months ago began to have trouble in walking. Also, pain in legs. Has not walked for three weeks. Has lost the feeling in legs for several months. Sometimes incontinence of bladder and rectum. Completely paralyzed in legs. Stroking the right sole causes marked extension of all the toes. Achilles jerks present on both sides. Knee-jerks excessive. Patellar clonus on both sides. Muscle-jerks are increased, and on passive movements there is marked spasm. Anæsthesia to simple touch to the upper third of thigh. No anæsthesia to deep pressure. She recognizes cold instantly, but warmth she does not recognize at all, or if it be very hot she recognizes it as cold. Some muscular wasting in both calves and both thighs. Spine is straight. No apparent deformities. There is a spot painful on deep pressure between the shoulder-blades.

A few days after her admission the following note is found upon her history:

Angle of mouth drawn somewhat to left. No paralyses of upper part of face, forehead, or eyes. Apparently no paralysis in arms. Patient lies all day in a semistupid condition, from which she can be aroused, but she will not obey either spoken requests or signs. She cries if she is subjected to questioning of any length. She has to be fed, for the reason that she refuses to feed herself. There is no difficulty in swallowing. There appears to be no paralyses of any of the external ocular muscles. She is somewhat emotional, sheds tears at times, especially when she is annoyed (when pins are stuck in her arm). After some difficulty the patient finally said that a pinprick hurt her (this after telling her that if she did not feel the pain the pin would be stuck through her arm).

The following notes were taken on June 6th, nine days after her entrance:

Patient now talks and obeys commands. Facial paralysis has developed. Bed-sore on buttock is observed this morning.

July 2d. For past two days patient has been complaining of pain in legs. She says she is unable to straighten her legs, although when examined they are found to be straight. For the past two days has had about five attacks, characterized by screaming and calling for someone to rub her arms. Her face twitches, becomes pale, then flushed. The arms and legs become rigid. Patient seems to be unconscious. The spell lasts three or four minutes. She is very emotional, crying at times. She died on July 12th.

Autopsy record discovers miliary tuberculosis in the left upper lobe of the lung, which is slightly adherent posteriorly, and on section is found to contain a cavity filled with purulent material. The rest of the upper lobe is studded with translucent nodules. Remainder of lung normal. Both lungs are slightly congested.

Kidneys cloudy and congested.

The surface of the liver is slightly mottled, and on section are seen areas of fatty metamorphosis around the common bile-duct.

Spleen normal.

Acute cystitis. The walls of the bladder are thickened and the mucous membrane is thickened and shows numerous hemorrhages. No pus. Genital tract shows nothing abnormal.

The pathological examination of the nervous system is as follows:

Sections are made at different levels of the spinal cord, of the medulla oblongata, and cerebral peduncles, and stained by Weigert's hæmatoxylin, Ehrlich's acid hæmatoxylin, and acid fuchsin.

Sections from the lumbar region show very marked degeneration of each crossed pyramidal tract. The degeneration at this level is almost confined to these tracts, except that there is a narrow zone of degeneration extending along the border of each anterolateral column. This is much narrower than the degeneration seen higher in the cord. Numerous holes are found in the posterior columns, and to some extent in the lateral columns, and are probably caused by dropping out of nerve fibres. There is an intense round-cell infiltration throughout the pia, the cells of which are mononuclear in type. There is also some round-cell infiltration about the vessels within the spinal cord, but at this level it is not very intense. The nerve cells of the anterior horns, so far as can be determined by the acid fuchsin stain, are not distinctly altered in the lumbar region.

Sections from different levels of the thoracic region present a zone of intense degeneration, extending around the entire periphery of the cord, and there are numerous small holes throughout the white matter of the posterior and anterolateral columns, caused by degeneration and dropping out of nerve fibres. The columns of Goll are not degenerated in the lower portions of the thoracic cord, but there is a very distinct degeneration of these columns in the upper portion of the thoracic region. The degeneration of the columns of Goll becomes much more intense as higher levels of the cord are examined, and the opposite is true for the degeneration of the crossed pyramidal tracts. In the latter tracts the degeneration is much more intense in the lumbar and lower thoracic regions than in the upper thoracic and cervical. Sections from the thoracic cord stained with a nuclear stain show intense round-cell infiltration about the vessels within the cord. There is also here, as elsewhere throughout the cord, an intense round-cell infiltration of the pia. Sections from the cervical region stained by Weigert's hæmatoxylin show a zone of intense degeneration extending around the entire periphery of the cord, and, as in the thoracic region, of nearly uniform thickness. (Fig. 1.) This is probably caused by the thickening of the bloodvessels extending from the periphery of the cord inward. The columns of Goll are much degenerated, though not completely so. The posterior and anterolateral columns show numerous holes, caused by the dropping out of nerve fibres. It is very likely that the degeneration along the periphery of the cord was the result, in part at least, of toxic substances conveyed by the arteria corona. Occasional proliferation of round cells, extending from the pia into the cord by means of the numerous septa, is found (Fig. 2); but these proliferations are not numerous. The crossed pyramidal tracts are not degenerated in the cervical region. The nerve cells of the anterior horns in the cervical region do not appear to be distinctly degenerated. Sections through the medulla oblongata, at the level of the lower olives, do not show degeneration.

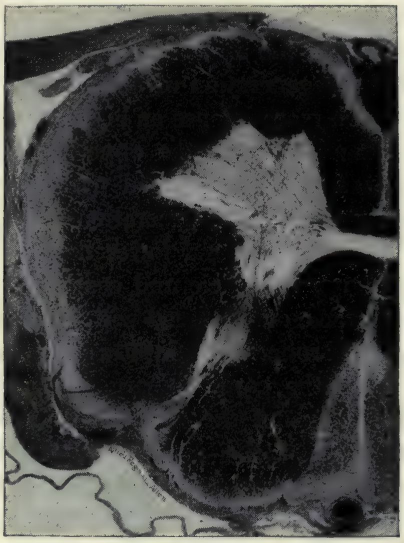

FIG. 1.—Section from the cervical region of the spinal cord stained by the Weigert hæmatoxylin method. A zone of degeneration of nearly uniform width extends around the periphery of the cord. The columns of Goll show secondary degeneration.

Sections through the cerebral peduncle show the same intense round-cell infiltration of the pia as seen throughout the spinal cord, and some of the vessels of the pia are much thickened. A round-cell infiltration is seen also about the blood-vessels within the cerebral peduncles, and in the pia over the peduncles and pons.

The case is one of intense meningoencephalomyelitis, probably of luetic origin, and presenting in the spinal cord the appearance of systemic disease, but not being, however, a true systemic disease. A tuberculous origin cannot be entirely excluded, but is not probable. The apparent systemic degeneration is evidently produced by disease of the bloodvessels within the cord, especially in the thoracic region. It is because of this that the column degeneration, as seen in the crossed pyramidal tracts and columns of Goll, increases in intensity in the direction in which these columns transmit impulses, that is to say, in the crossed pyramidal tract greater in the lower part of the cord, and in the columns of Goll greater in the upper part.

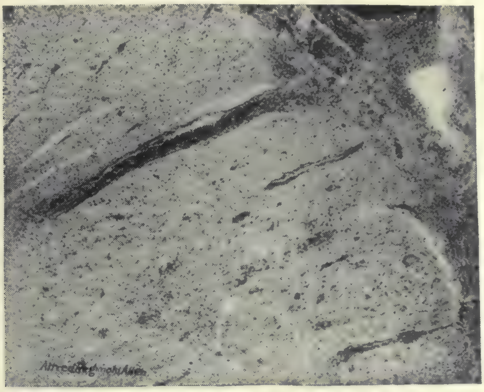

FIG. 2.—Round-cell infiltration of pia and about centripetal arteries in the peripheral zone of degeneration. Section from the thoracic region.

This case is of particular interest on account of the intense annular degeneration.

Pierre Marie[1] says that a large number of cases of combined posterolateral sclerosis are of vascular origin. R. T. Williamson[1] is the author of an extensive monograph designed to show the vascular etiology of system disease in general. In the case I am reporting the vascular etiology seems to me most apparent. In arguing for the vascular origin of posterolateral sclerosis Pierre Marie says: "The fact of the direct pyramidal tract being unaffected, while the crossed pyramidal is badly affected by sclerosis affords, as I have already said, one of the best arguments which can be used in opposition to the theory that this form of sclerosis is of systemic nature." In view of the difference in symptomatology, it is interesting to note that the case here reported presents pathologically, with the exception of round-cell infiltration, a picture very analogous to Friedreich's ataxia. In considering the probability of intoxication from the bloodvessels, one should not lose sight of the fact that the spinal cord receives both anteriorly and posteriorly blood from the same source, namely, the vertebral arteries. Now, if we are to ascribe vascular morbidity as the cause of distinct system disease, I ask why, if the inflammatory and degenerative process is due to an intoxication in which the bloodvessels are first affected, and, secondly, although practically synchronously, the adjacent tissue, why, I say, does the process seem so selective in these system cases? Dr. Williamson's paper, mentioned above, would lead us to look upon the following diseases as entirely vascular in origin. Kindly note how selective the morbid process must be in some of them: Poliomyelitis, Friedreich's disease, combined posterolateral sclerosis, acute anterior poliomyelitis of the infant (infantile paralysis), acute anterior poliomyelitis of the adult, subacute and chronic poliomyelitis, disseminated poliomyelitis, acute transverse poliomyelitis, disseminated sclerosis, locomotor ataxia, and paralysis agitans.

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[1] Pierre Marie. Leçons sur les maladies de la moelle.

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[1] On the Relation of Diseases of the Spinal Cord to the Distribution and Lesions of the Spinal Bloodvessels.

In closing, I would like to call attention to the points in common between the case I am reporting and Erb's syphilitic spinal paralysis, the latter showing a spastic paralysis of the legs, with the consequent exaggeration of reflexes and spastic gait, also disorder of sensibility and incontinence of urine, slight muscular contractions, and chronic course.

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BULBAR SYMPTOMS OCCURRING WITH CARCINOMA OF PARTS OTHER THAN THE NERVOUS SYSTEM, AND RESULTING FROM INTOXICATION.

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(From the Neuropathological Laboratory of the University of Pennsylvania.)

It is well known that cerebral symptoms may occur in the course of carcinoma of the breast, stomach, œsophagus, or of any of the internal organs. Among the conditions that have been reported are coma, mental apathy, or other psychical disturbances. Recently I had under my care in the Home for Incurables a patient with carcinoma of the breast, who for some time before her death had delusions of varying character, and was with difficulty restrained from jumping out of the window and from disrobing at improper times.

Hemiplegia, with or without aphasia, monoplegia, aphasia, convulsions, either general or Jacksonian in type, pontile symptoms, as facial paralysis on one side, and hemiparesis upon the other, symptoms of involvement of the different cranial nerves, and bulbar phenomena have all been recorded as occurring in the course of carcinoma. These, however, are by no means common.

Spinal symptoms are still more rare. Nonne recorded an instance of spasticity of the lower limbs occurring in the course of carcinoma of the uterus, in which on microscopic examination a degeneration of both pyramidal tracts with partial degeneration of the tracts of Gowers and of the direct cerebellar tracts was found. There were no signs of cancer anywhere in the brain or cord. Of course, the occurrence of metastatic tumors either in the brain or cord is well recognized.

Bulbar symptoms as a complication of carcinoma have been previously mentioned, so far as I know, only by Bruns and by Spiller and myself.

In our report, published in May, 1904, we reviewed the literature upon the subject, but it might be well to repeat the more important points.

In Bruns' case, which was like ours, one of vertebral carcinomatosis, the bulbar symptoms consisted in a right-sided atrophy and paralysis of the tongue, right-sided paralysis of the soft palate, difficulty in swallowing, increase of the pulse rate, and occasional vomiting. The atrophy and paralysis of the tongue later became bilateral.

In our case the patient was in such a low condition when she was examined that the only distinct bulbar symptoms we could elicit were a bilateral atrophy of the tongue and a distinct inability on her part to protrude her tongue much beyond her teeth. There was also an atrophy of the face and lips, but this may have been a part of the general emaciation.

Bulbar symptoms due to malignant tumors other than carcinoma have been recorded by Nonne, Oppenheim, and Senator. In all of these cases the tumor was a lymphosarcoma, and invaded respectively the spinal cord, chest, and ribs. In none of these cases did the microscopic examination reveal the cause of the bulbar phenomena.

I have had under my care in the Philadelphia Home for Incurables a woman, aged fifty-nine years, who for thirteen years before her death had a carcinoma of the right breast. She was operated upon seven and again five years ago, part of the growth being removed each time. One year ago her right arm began to swell. When I first examined her, which was two months before her death, the cancer of the right breast had extended into the chest, the glands of the neck were involved, and the right upper limb was extremely swollen. There were no symptoms of gastric metastasis. Unfortunately her throat and larynx were not examined.

Five months before her death the patient first complained of difficulty in swallowing, and also noticed that the voice was hoarse. Since that time articulation gradually became more difficult, until finally she could not talk above a whisper. When I examined her she could swallow only fluids and with difficulty, except that at extremely rare intervals she was able to swallow hard substances. Speaking fatigued her. There was no dribbling of saliva. The tongue was protruded straight, and there was no tremor or atrophy. All of the movements of the tongue were well performed. The masseter contracted firmly on either side. There was no weakness in the distribution of either seventh nerve. The pupils were normal, and responded promptly to light, accommodation, and convergence. The extraocular movements were normal.

We were allowed to remove only the brain at the autopsy. No gross macroscopic lesions were found, except in the cerebellum, where some of the bloodvessels were extremely congested. In a microscopic examination no round-cell infiltration was apparent around these bloodvessel walls.

The nerve cells of the paracentral lobule of each side, of the cerebellum, and the nerve cells of the nuclei of the sixth, seventh, ninth, tenth, and twelfth nerves were examined by the thionin stain.

The nerve cells of the paracentral lobule, as shown by the Betz cells, were normal on each side. The cells of Purkinje in the cerebellum were not altered. The cells of the nuclei of each hypoglossus nerve were also found in a normal condition, but some of the nerve cells of the nucleus ambiguus and of the dorsal vagus nucleus, both in its ventral and dorsal parts, were distinctly diseased. Some of the nerve cells of the glossopharyngeus nucleus were also altered. In the nuclei of each seventh and sixth nerves some of the nerve cells were distinctly altered.

This alteration consisted in a displacement of the nucleus to the periphery, and in disintegration of the chromophylic elements. There was no increase of yellow pigment, and no swelling of the cells. The pial covering both on the cortex and on the base of the brain showed no carcinoma cells.

Several explanations have been advanced to account for the various cerebral symptoms present with carcinoma. In a discussion of this subject before the Biological Society of Hamburg, in January, 1900, Saenger expressed himself of the opinion that the various phenomena found in this disease were not due to a toxic cause, as Oppenheim and Bettelheim thought, but that they were due to definite organic changes, basing his opinion upon the finding of carcinoma cells in the cortical and basal pia.

He rightfully opposed Oppenheim's views, because this author thought that the toxic changes were localized to certain parts of the brain.

Nonne, in the discussion, expressed the view that while some of the symptoms may be explained by a metastasis, most of the macroscopic findings were negative, and he inclined to the toxic theory. He quoted many cases of his own, of hemiplegia with or without aphasia, of convulsions, etc., in which, on microscopic examination, no changes were found. Saenger, in reply, remarked that these cases were insufficiently studied.

Recently, Hudovernig, in a case of carcinoma of the œsophagus, found, by examination of the medulla oblongata, changes in some of the nerve cells of the nucleus ambiguus, and of the dorsal vagus nucleus. Most of the alterations, which were studied by Nissl's method, were in the so-called dorsal vagus nucleus.

He concludes that as the œsophagus is innervated by the vagus nerve, especially by its sensory fibers, these changes illustrate the so-called reaction at distance. He quotes Parhon, who found changes in the nerve cells of the hypoglossus nucleus due to carcinoma of the tongue.

In the case reported by Spiller and myself alterations in the cells of each hypoglossus nucleus were found, in our case there was no carcinoma of the tongue, but atrophy of either side.

The study of the case I now report shows that as a result of carcinoma we may have alterations in the nerve cells of the different bulbar nuclei, and that these are probably toxic in origin. The failure to examine the throat does not exclude an involvement of the recurrent laryngeal nerve, but granting, for the sake of argument, that this did occur, how are we to explain the changes in the nerve cells of the sixth and seventh nuclei except by toxic alteration.

It is recognized that certain poisons, as, for instance, those of uræmia, may produce definite changes in the nerve cells not only of the medulla oblongata, but also of other parts of the brain. Such alterations were described by me last year. In those cases and in the present one the changes were not confined to one group of cells, but were symmetrical, opposing the views advanced by Oppenheim. They may, however, be more marked on one side, as, for instance, in my case of uræmic hemiplegia, in which the alterations were so intense that a resulting primary degeneration of the corresponding motor tract was found.

The assumption of Parhon and of Hudovernig is probably not correct, inasmuch as these alterations were found only in a very few cells. In Hudovernig's paper no mention is made of the condition of other nuclei than the twelfth; the latter were normal.

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FIBROUS NODULES IN THE CEREBRAL PIA-ARACHNOID, CAUSING THE APPEARANCE OF TUBERCULOUS MENINGITIS.

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I WISH to report a case of interest because of the peculiar nodular formations in the cerebral pia-arachnoid. The pathological material upon which this paper is based was placed at my disposal by Dr. William G. Spiller.

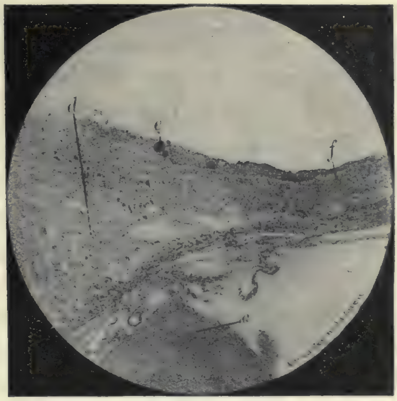
The clinical history, taken from the records at the Philadelphia General Hospital, is very incomplete. The man was sixty-four years of age at his death, and had been a peddler by occupation. The family history revealed nothing of importance and although he had been a heavy drinker, his health previous to his last illness had been good During the eight months before his admission to the hospital he had occasional attacks of dyspnœa, accompanied by œdema of the legs. These attacks were much more frequent in the latter part of this period. On admission to the hospital he complained of headache, vertigo, backache, weakness, and abdominal pain. When examined he was found to have a large mass in the abdomen and an enlarged liver. He was a rachitic dwarf, only four feet tall, with a rachitic head and chest.

Death occurred about three months after his admission to the hospital, and at autopsy there was found a large retroperitoneal sarcoma with extensive metastasis to the liver, lungs, and kidneys.

The pia covering the convexity of the brain is studded with small white nodules, which at the necropsy were supposed to be sarcomatous on account of the coincident abdominal growth. These nodules are found generally distributed over the entire surface of the cerebral hemispheres, but do not seem to be present in the basal parts. They vary considerably in size, but on an average are about as large as the head of a small pin. They are found especially in the pia covering the fissures, and are usually close to a bloodvessel, although a few do not show such intimate connection. Some very slight thickenings of the pia, about the size of the point of a pin, are found upon the surface of the convolutions; these are always much smaller than those in the fissures. The nodules are white and opaque, and resemble very closely those occurring in tuberculous meningitis, so closely, indeed, that a differential diagnosis by the unaided eye would be impossible.

The Pacchionian bodies are numerous and very large.

On histological examination the nodules are found to be directly within the substance of the pia mater, and consist of wavy, fibrous, connective tissue like that of the pia, but containing very few nuclei, and in this respect differ greatly from the pia; also the bands of connective tissue are thicker in the nodules than they are in the pia. Over the outer surface of a nodule and extending somewhat along the outer border of the adjacent pia there is a considerable proliferation of the endothelial cells like that described by Dr. W. G. Spiller[1] in a case of lead poisoning with encephalopathy. The nodules are not more adherent to the cortex than is any other part of the pia, and have none of the appearance of tubercles when examined microscopically.



Fibrous nodule in the pia-arachnoid. *d*, fibrous tissue poor in nuclei; *e*, corpora arenacea; *f*, thickened pia; *c*, cerebral cortex.

In the proliferated endothelial cells upon the upper part of these nodules are found numerous corpora arenacea. It is worthy of note that these bodies are found in the proliferated endothelial cells. There is no round-cell infiltration in the cortex.

Some of the Betz cells in the paracentral lobules show an intense, yellow, granular pigmentation.

The pia is thickened in some places where there are no nodules, as over the left paracentral lobule; and the pia, where thickened, has the same histological structure as the fibrous nodules.

In order to make clear the pathological change occurring in the pia it will be necessary first to discuss briefly the normal histology of the membrane.

The early teaching was that there are two distinct membranes—an outer, delicate, non-vascular layer of fibrous tissue which bridges the sulci without dipping into them, and an inner, vascular membrane which closely invests the entire surface. Between the two there was said to be a considerable space traversed by numerous trabeculæ.

Dr. Batty Tuke[2] dissents from this opinion. He holds that the pia and arachnoid should be looked upon as only one membrane, of which the arachnoid is merely the outer layer, and that the bloodvessels are distributed between the two layers instead of within the layer. W. F. Robertson[3] published in 1895 a careful study of the pia-arachnoid, in which he comes to the conclusion that it is but one membrane, lined both externally and internally with endothelium, and composed of a meshed connective tissue the spaces of which are also lined with endothelium. These spaces are larger at the centre than at the edges, this giving the appearance of layers, which led to the belief that there are two separate membranes. The same arrangement of trabeculæ and intervening lymph spaces is maintained throughout the membrane, and thus it has the structure of a spongy lymph sac. The bloodvessels occur in any part, but the veins especially occur near the outer endothelial surface. The capillaries and arterioles are chiefly in the deeper parts, but may occasionally be seen near the outer surface. On account of the scarcity of capillaries it is evident that they are but little, if at all, required for the nutrition of the membrane, which seems to be maintained by the cerebrospinal fluid. The arterioles of the pia-arachnoid have no proper adventitial coat, no layer of longitudinally disposed fibrous tissue such as is found in vessels elsewhere. They are, as it were, naked vessels surrounded by trabecula and lymph spaces. Middlemas and Robertson[4] considered that the "whole extra-vascular structure of the soft membranes may be looked upon as the conjoined and hypertrophied adventitial coats of the pial vessels."

Of the pathological changes occurring in the pia there are two classes, the inflammatory and the non-inflammatory. The most common of the non-inflammatory conditions is the cloudiness and thickening observed in senility and in the insane. Various opinions have been expressed as to the etiology of this change. Bayle[5] thought these alterations are due to a chronic meningitis. Bevan Lewis[6] is of the opinion that when they are very intense we must infer an inflammatory agent; but when they are less marked, especially in senile atrophy of the brain, they may result from other causes than inflammatory action. The condition that Ziegler[7] terms "chronic arachnitis" is due, he says, to fibrous thickening, endothelial hyperplasia, and more rarely to cellular infiltration. Robertson[3] considers that in typical cases these changes are due to a slow hyperplasia of the connective tissue, and to marked proliferative and degenerative changes in the endothelial cells lining the arachnoid spaces, and in those of the outer surface. He found the signs of inflammation to be present only in cases of advanced general paralysis, syphilitic insanity, and in the condition known as "purulent infiltration of the pia-arachnoid." Robertson also speaks of the fact that hypertrophy of the Pacchionian bodies frequently accompanies this condition of pial thickening.

Obersteiner[8] describes the non-inflammatory changes in the pia-arachnoid as follows: "In the brains of many adults, especially, however, in those advanced in years, small, light, cloudy areas are found scattered over the arachnoid. These are seen over the convexity of the brain near the middle line, and over the bloodvesels of the pia. The arachnoid becomes thickened and cloudy in consequence of chronic irritative processes. This condition is frequently found in insanity and alcoholism, but also occurs in simple atrophy of the brain, namely, senile atrophy.

"Different from these are small, disseminated, white lesions that are distributed regularly over the entire convexity, and are formed also by thickening of the arachnoid. They are found in cases of insanity, usually in dementia.

"Numerous small proliferations of the overlying endothelium may be found in the beginning of paralytic dementia, giving the arachnoid a roughened appearance."

The condition observed in my case is undoubtedly the same as that described by Obersteiner as "small, disseminated white lesions," but I do not find any reference to them by Stroebe,[9] or by Sacki and Schmaus[10] in their recent book on the pathology of the nervous system.

Microscopically the nodules conform very closely to the structure of the Pacchionian bodies, and differ from them in location and in the absence of a pedicle.

A description by Fuchs[11] of the Pacchionian bodies is as follows: "The Pacchionian granulations break through the inner layer (of the dura) on both sides of the longitudinal sinus and enter, with a thin pedicle, the parasinoidal spaces. The pedicle and the greater part of the villus, which forms a swelling on the inner side of the parasinoidal space, are composed of a loose, fine-fibred tissue, very poor in nuclei, but presenting all the characteristics of connective tissue. The connective-tissue fibres run parallel in the pedicle, but radiate out in the villus. In the villus the tissue forms a very fine-meshed reticulum, in the spaces of which frequently small hemorrhages are found. Capillaries are also found in the stroma of the villus. The central part of the villus contains few nuclei, and appears necrotic, but closer examination shows it to be intact, and the nuclei of the tissue may be seen near the periphery. The nuclei are small and are most numerous at the base of the villus."

As will be seen from the foregoing description, the microscopic appearance of the nodules in the case I have described corresponds very closely with the appearance of a Pacchionian body, but the distribution of the former over the entire convexity of the brain, and their mode of development —viz., instead of being an outgrowth of the pia-arachnoid they are fairly within the membrane, and especially in the inner portion—mark them as distinctly pathological.

The etiology of the nodules in this case is not clear. There were no signs of inflammation and there were no symptoms of mental disturbance noted. Outside of a history of alcoholism there is no apparent cause for the condition.

An interesting and very practical pathological observation upon this case is the exceedingly close resemblance to tuberculous meningitis when the examination is made by the unaided eye. The nodules are in every respect the exact counterpart of miliary tubercles in size, appearance, number, and distribution. Microscopically, however, they are entirely different, as instead of the typical cellular arrangement of a tubercle we have a fibrous tissue that is very poor in cells. As many pathological diagnoses are made without histological examination, the mistake of calling this nodular condition a tuberculous meningitis might readily be made.

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PRIMARY DEGENERATION OF THE PYRAMIDAL TRACTS; A STUDY OF EIGHT CASES WITH NECROPSY.

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Amyotrophic lateral sclerosis is regarded as an uncommon disease of the nervous system, but the opportunity has been given me in the last six years to study eight cases of primary degeneration of the central motor tracts, with necropsy. In six of these cases degeneration of the nerve cells of the anterior horns was present, so that these were cases of amyotrophic lateral sclerosis. Four of the cases have been reported, two by myself alone and two in association with others. One of the remaining four cases has been referred to briefly.

Two of these eight cases were ascending and at first unilateral in type, and later the paralysis became a triplegia (Cases IV. and VIII.). One began as a hemiplegia, and later the other side of the body become affected (Case V.). Two may be regarded as uncomplicated cases of primary degeneration of the pyramidal tracts without implication of the nerve cells of the anterior horns of the spinal cord, and in neither of these cases was muscular atrophy present (Cases IV. and VI.). In one case only could degeneration be traced as high as the motor cortex (Case II.), but in two it could be traced into the internal capsule (Cases IV. and VII.), and in two into the cerebral peduncle (Cases V. and VIII.). In three of the cases the degeneration extended as high as the pons and not beyond (Cases I., III., and VI.).

Having had, therefore, within six years the opportunity to study eight cases of primary degeneration of the pyramidal tracts with necropsy, of which six could be regarded as examples of amyotrophic lateral sclerosis, and having seen within this time a number of clinical cases of this type, I am obliged to conclude that amyotrophic lateral sclerosis is not so rare a disease in my experience as in the experience of some others.

It is my purpose to discuss the four unreported cases and to refer to those already reported in reference to certain aspects of the disease. It has seemed to me well to give brief abstracts of the four cases already reported:

CASE I. (Dercum and Spiller).[1]—The patient was a male, aged fifty-three years. He was admitted to the Philadelphia General Hospital on June 14, 1892. For five or six months before admission he had complained of pains in both lower limbs below the knees, and probably these were the pains of fatigue, as he would become very tired by night. On admission the lower limbs were somewhat spastic and the reflexes were exaggerated. Cutaneous sensibility was not altered. In the summer of 1895 weakness and rigidity of the lower limbs had become so great that he was confined to a wheel-chair. About this time speech was noticed to be of a bulbar character. The upper limbs gradually became spastic. By April, 1897, he had become bedridden and had occasional retention of urine and loss of control over his bowels. He died on January 7, 1898, after his limbs had become weak, spastic, and atrophied, and after the tendon reflexes had become exaggerated and bulbar symptoms had developed.

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[1] Journal of Nervous and Mental Disease, February, 1899

The microscopic study revealed degeneration of the nerve cells of the anterior horns of the cervical and lumbar regions, rarefaction of the anterior horns, degeneration of the crossed and direct pyramidal tracts, in the former extending beyond the area of these tracts, and extending as high as the pons, and slight sclerosis of the posterior columns in the lower cervical and upper thoracic regions.

Case II. (a patient in the service of Dr. Mills at the Philadelphia General Hospital).[1]—A man, aged fifty-five years, contracted a severe cold in October or November, 1897. He said he had been healthy until that time. Toward the end of 1897 he began to have dysphasia, involuntary dribbling of saliva, and loss of power in the upper and lower limbs. These symptoms increased until he became confined to his bed, and almost completely paralyzed. The deep reflexes were exaggerated, and fibrillary tremors were noticed in the tongue and other muscles. Sensation was not disturbed. Muscular atrophy was intense. Death occurred on November 17, 1898.

The disease began with bulbar symptoms and involvement of the limbs, and ran a rapid course, the patient dying about a year after his symptoms first became distinctly perceptible.

The muscles of the upper extremity, examined microscopically, were considerably atrophied, and those near the distal end of the limb were more altered than those near the proximal end. In an interosseous muscle examined the atrophy was found in an extreme degree.

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[1] Contributions from the William Pepper Laboratory of Clinical Medicine, 1900.

The ulnar nerve was found to be partially degenerated.

The anterior roots of the spinal cord were atrophied.

The nerve cells of the anterior horns were degenerated, especially in the cervical region. Degeneration in the motor tracts extended from the cerebral cortex to the lumbar region of the spinal cord.

Some of the motor bulbar nuclei were diseased. The corpus callosum was much degenerated by the Marchi method. The giant cells of the paracentral lobule were numerous but much pigmented.

Case III. (a case in my service in the Philadelphia General Hospital).[1]—A woman, aged fifty years, developed suddenly weakness in her left upper limb, with loss of speech. The power of speech was regained after a day or two, but was never again normal. The weakness almost entirely disappeared from the left upper limb after about three weeks. Two years after this attack she noticed that she was weak in her lower limbs, and the weakness gradually increased so that walking became difficult. The reflexes in all the extremities were exaggerated, and the Babinski reflex was obtained. The gait was not decidedly spastic. The right upper limb was not distinctly paretic, but the left was a little weak. No objective sensory disturbances were found, and no pains were felt in the limbs. Muscular atrophy was not observed, although the palsy of the lower limbs had existed more than a year. The reaction of the irides was sluggish. The left side of the face was slightly paretic, but the right side also probably was not normal. The patient had been constipated and had noticed an increasing difficulty in retaining the urine.

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[1] Journal of Nervous and Mental Disease, May, 1902.

Degeneration of the pyramidal tracts was found extending as high as the pons, but not above this. This degeneration was less intense in the anterior pyramids than in the spinal cord, and was equal on the two sides of the cord. The other tracts in the cord were normal. No meningitis was present. The cells of the anterior horns of the spinal cord were in part diseased, and the nuclei of the hypoglossal nerves were probably not absolutely normal.

CASE IV. (Mills and Spiller).[1]—The patient, a male, aged sixty years, was in the service of Dr. Mills at the Philadelphia General Hospital. Hemiplegia gradually developed on the right side, the lower extremity being more markedly and for this reason probably earlier affected than the upper; the case, therefore, at first belonging to the clincal type of unilateral progressive ascending paralysis. After several years the left lower extremity also became paralyzed, but not to the same extent as the right. The reflexes were all markedly exaggerated, the Babinski response being present. Sensory symptoms were absent.

Microscopic examination showed intense and long-standing degeneration of the right crossed and of the left direct pyramidal tracts, the degeneration extending into the pons, but not into the left cerebral peduncle; also comparatively recent degeneration of the left crossed and of the right direct pyramidal tracts, traced by the method of Marchi into the lower part of the right internal capsule. The nerve cells of the anterior horns of the lumbar cord were not distinctly altered. They were numerous and some were much pigmented, which, considering the man's age, was not remarkable. The chromophilic elements were well formed and the nuclei central. It may be said that the nerve cells of the anterior horns of the lumbar region had the appearance usually seen in persons of the same age. The nerve cells of the anterior horns of the cervical region were normal, although in examining a number of sections a few cells were found that were not normal, but the nerve cells in general were remarkable for their excellent state of preservation. No lesions, degenerative or focal, were found elsewhere in the brain or spinal cord; the case, therefore, was one of primary degeneration of the motor tracts, much greater and older in the right crossed and left direct pyramidal tracts.

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[1] Journal of Nervous and Mental Disease, 1903, p. 385.

CASE V. Kate D. (a case in my service at the Philadelphia General Hospital).—The patient, a woman of middle age, was admitted to the hospital on March 27, 1901. Notes taken at the time of her admission state that about one year previously she became paralyzed in the right side of her body and face, and that later her left upper and lower limbs became paralyzed, and she lost the power to articulate. The pupils were equal, and the reaction to light in accommodation and in convergence was preserved. Sensation was normal. She had not had any miscarriages.

Notes taken by me on June 14, 1901, are as follows:

During the examination the patient makes no sound, and apparently is unable to speak at all; she is extremely emaciated. The wasting is especially marked in the small muscles of each hand. The thenar eminence is entirely flattened, so that the hand has the simian form. The fingers of the left hand are flexed at the phalangeal articulations; the little and ring finger being more flexed than the middle and forefinger, and the fingers are turned toward the ulnar side. The right hand is as much wasted as the left, and the fingers are flexed at the phalangeal articulations, but all the fingers seem to be equally flexed, and they are also drawn toward the ulnar side. The forearms and upper arms are very much wasted, the forearms being more wasted than the upper arms. The only contractures present are in the fingers of each hand, where there is tendency to flexor contracture, but the contractures are not very pronounced. Fibrillary tremors are present in the upper arm on each side, and the little finger of the right hand is jerked involuntarily occasionally.

The right and left biceps tendon reflexes are exaggerated, as are also the wrist reflexes.

The muscles are so wasted that the reflex movements are not very pronounced, but there is no doubt that all the reflexes are increased. Considerable spasticity is present in the upper limbs at all parts.

The face is also emaciated, though not extremely so; the lips are much wasted, and the patient is unable to blow out a lighted match. When she laughs or cries the corners of the mouth are drawn up, equally and well. The eyelids can be closed, and are held quite firmly shut.

The tongue is extremely atrophied, and the patient is entirely unable to protrude it.

The soft palate moves slightly, and appears to be paretic.

Heart sounds are loud and regular, and there are no murmurs.

Tactile and pain sensations in the upper limbs, chest, and face are normal.

The intelligence of the patient is very good; she nods her head correctly in answer to questions.

Lower limbs are also much wasted, but not so much proportionately as the upper.

The feet are contractured in the position of talipes equino-varus, and the contractures are pronounced.

The toes of each foot are contractured in flexion. Spasticity is very marked in the lower limbs, and when either limb is lifted from the bed the leg is firmly extended at the knee, but by gentle and gradual pressure the rigidity can be overcome.

Emaciation in the lower limbs is general, and is not more marked in the soles of the feet than elsewhere in the limbs.

The patellar reflexes are exaggerated. The Achilles jerks are not obtainable, because of the permanent contractures.

The Babinski reflex is peculiar; irritation of the sole of either foot causes slight extension of all the toes, except of the great toe, which does not move. The extension is slight, because of the contracture of the toes in flexion.

Sensation for touch and pain in the lower limbs is normal.

Diagnosis. Amyotrophic lateralsc lerosis with bulbar involvement.

The notes of an examination of the throat and larynx by Dr. M. Marshall are as follows: The patient has complete paralysis of the tongue and partial paralysis of the vocal cords, this being more marked on the left side. The soft palate is not involved.

A cough developed slowly, difficulty in swallowing became gradually more intense, rales were heard throughout the lungs, and the heart sounds were weak. She died on June 28, 1901, having grown progressively weaker, and having presented symptoms of the disease a little more than a year.

The necropsy was made by Dr. W. F. Hendrickson, and his pathological diagnosis was:

Acute bronchopneumonia, chronic ulcerative pulmonary tuberculosis (apex), œdema of pia-arachnoid of the brain, atrophy of cerebral convolutions, slight hydrocephalus, acute pleurisy, early acute splenic tumor, chronic diffuse nephritis, general enteroptosis.

The result of my microscopic examination is as follows:

Lumbar Region. The anterior and posterior roots stain well. The nerve cells of the anterior horns are few in number, some are very small and contain few chromophilic elements and are much pigmented. The crossed pyramidal tracts are much degenerated and equally so on the two sides. The right direct pyramidal tract may be a little less deeply stained than the surrounding tissue. The anterior horns are well stained by the Weigert hæmatoxylin method. The Marchi method shows a few black dots scattered over the anterolateral columns, but not more in the pyramidal tracts than elsewhere.

Thoracic Region. Both direct and both crossed pyramidal tracts are much and symmetrically degenerated. The posterior columns are not affected. The anterior and posterior roots stain well. Very marked recent degeneration is shown by the Marchi method in both direct and both crossed pyramidal tracts, and slighter degeneration throughout the anterolateral columns. The recent degeneration is more intense in the right direct pyramidal tract than in the left, and about equal in the crossed pyramidal tracts.

Lower Cervical Region. Pronounced degeneration of both crossed and of both direct pyramidal tracts is shown by the Weigert hæmatoxylin method, but the posterior columns are normal.

The anterior horns are not so deeply stained as the posterior. The direct and crossed pyramidal tracts show recent degeneration by the Marchi method, and the degeneration is more intense in the right direct and the left crossed pyramidal tracts; the degeneration appears to be of longer duration in the right crossed and the left direct pyramidal tracts. A slight recent degeneration is found throughout the anterolateral columns. Very few nerve cells are found in the anterior horns and those that are present are much degenerated.

Medulla Oblongata. The nucleus of each twelfth nerve is completely degenerated, and there are no intramedullary fibres of either nerve. No nerve cells are found in either twelfth nucleus. The anterior pyramids are partially and equally degenerated. Nerve cells are found in each posterior nucleus of the vagus, but in many the nuclei are eccentric. The intramedullary fibres of each ninth and tenth nerve and each nucleus ambiguus are much degenerated. The intramedullary portion of the sixth nerve stains well, but the seventh nerves and their nuclei are much degenerated. The pyramidal tracts of the lower part of the pons are very slightly degenerated.

Cerebral Peduncle. The middle portion of the foot of one cerebral peduncle shows numerous small holes from which nerve fibres have fallen out, and this area by the Marchi method shows a few black dots, otherwise the foot of the peduncle is normal.

Recent degeneration, as shown by the Marchi method, is unimportant in the left paracentral lobule.

Case VI.—William B., aged forty-two years, was admitted to the Philadelphia General Hospital on April 5, 1899. At that time he was in the service of Dr. Edward Martin on account of an ulcer of the leg, which he had had about four years. He was not very intelligent. The patellar reflexes at that time were slightly exaggerated. He was cured of the ulcer.

He came into my service in the same hospital, and on August 24, 1903, the following notes were dictated by me:

The man says he has been paralyzed in the lower limbs about four years and five months; the paralysis developed slowly. He has not had any pain in his limbs. He acknowledges having drunk freely beer and porter. There is no history of injury. The lower limbs are exceedingly spastic and equally so. The voluntary movements in the lower limbs are almost entirely confined to the muscles about the hips, and he has a little more power in the left lower limb than in the right. He is unable to flex either leg upon the thigh or either foot upon the leg, and has marked talipes equinus on each side. The toes are dorsally flexed at the metatarsophalangeal articulations, and extended at the other phalangeal articulations. He has an ulcer on the inner side of the right ankle. The limbs are not wasted. The spasticity of the lower limbs is so great that it is impossible to obtain any movement of the legs upon the thighs in testing the patellar reflex, but distinct, though not intense, contraction is seen in the quadriceps muscles. Ankle clonus and the Achilles reflex are prevented on each side because of contractures. Babinski's reflex is present on each side, but is weak because of the contracture of the toes.

Sensation for touch and pain seems normal in the lower limbs. He has incontinence of urine and feces.

Voluntary power in the upper limbs is normal. The movements of the upper limbs are somewhat ataxic. The biceps tendon and triceps tendon and wrist reflexes are about normal on each side. The upper limbs, including the hands, are not atrophied. He has no girdle sensation.

The pupils are about equal. The iritic response in each eye is prompt to light, in accommodation and in convergence. The movements of the eyeballs are normal in all directions. The tongue deviates slightly to the right, and may be slightly atrophied. The movements of the facial muscles are normal. The masseter contracts firmly on each side.

Diagnosis. Spastic paraplegia. Death occurred on September 5, 1903.

The pathological diagnosis was: Chronic parenchymatous nephritis, fatty infiltration of liver, and congestion of the lungs.

The result of my microscopic examination is as follows:

Lumbar Region. Each crossed pyramidal tract is partially degenerated, and the degeneration is confined to these tracts. The posterior and anterior roots are normal. The nerve cells of the anterior horns are in a fair state of preservation, or at least their alteration is slight.

Midthoracic Region. The crossed and direct pyramidal tracts are distinctly degenerated. There is a slight degeneration in the area of Gowers' tract on each side. The anterior and posterior roots are normal.

Lower Cervical Region. The crossed and direct pyramidal tracts are much degenerated, but the degeneration is not complete, and, therefore, is like that of primary degeneration. The anterior and posterior roots are normal. The nerve cells of the anterior horns are in a very fair state of preservation; some may not be normal, but the alteration is insignificant. There is a slight sclerosis of the columns of Goll in the cervical region. Meningitis is not found anywhere.

Medulla Oblongata. The anterior pyramids are almost normal.

Case VII. has been referred to very briefly. The clinical and necropsic notes as obtained from Dr. W. E. Robertson[1] are as follows:

Sarah L., aged about sixty-two years, was admitted to the Episcopal Hospital, Harrison wing for chronic diseases, with a diagnosis of chronic rheumatism. At that time she was able to get about on crutches, but her joints were stiff, and her hands were deflected to the ulnar side, showing the chronicity of the trouble. Her mind was clear, and, except as stated, she resembled any other woman of her years. Gradually some change in her gait was noticed, and she became at first somewhat spastic. At this time sugar was present in her urine, although it almost disappeared under appropriate dietetic treatment. Soon a change in the character of her voice was noticed, and, shortly following, some difficulty in swallowing food. She often choked in swallowing. This choking was progressive, and with it paralysis of all four extremities came on, so that she became totally helpless and eventually lost control of bladder and bowel. Death resulted from exhaustion superinduced by the inability to take food.

Necropsy five hours after death. Body of a small woman. No lividity or rigidity. Subcutaneous fat well preserved.

Thoracic Cavity. A few adhesions on both sides. No apical scars. Left lung crepitated throughout. Both lower lobes of the right lung were the seat of a croupous pneumonia. Heart small. Some thickening of all the valves, and in the mitral and aortic valves some atheroma. Aorta dilated just above semilunar valves.

Abdominal Cavity. Stomach fetal in type, retort-shaped, and vertical. Liver small, much congested, cirrhotic, and slightly fatty. Gall-bladder contained three large and two small stones. Common duct patulous. Spleen normal in size and consistency. Kidneys small and rather pale, firmer than normal. On section, cortex narrow; capsule striped with difficulty, tearing parenchyma. Pancreas apparently normal. Appendages and uterus normal.

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[1] Spiller, Robertson, and Wadsworth. The University of Pennsylvania Medical Bulletin, June, 1901

Brain. Meninges somewhat congested. There seemed also to be an excess of cerebrospinal fluid. Situated on the corpora quadrigemina was a firm tumor. It was round, about one inch in diameter, and was a fibroma. It had probably caused no symptoms, although it had considerably compressed one lateral lobe of the cerebellum.

The result of my microscopic examination is as follows:

Lumbar Region. The anterior and posterior roots stain well. There are fewer nerve cells than normal in the anterior horns, and many of those that are present are much degenerated. The crossed pyramidal tracts are degenerated and equally so.

Thoracic Region. The crossed pyramidal tracts are much degenerated, and one as much so as the other. The posterior columns are not degenerated.

The cervical region was not obtained.

Medulla Oblongata. The anterior pyramids are partially degenerated. The intramedullary fibres of the hypoglossal nerves are not so well stained as normally, and the nuclei of these nerves contain few cells. The nucleus ambiguus is degenerated on each side. The nuclei of the facial nerves do not contain the usual number of cells, and many of the cells of these nuclei are much degenerated. Much recent degeneration is shown by the Marchi method in each anterior pyramid.

Sections through one of the cerebral peduncles show much recent degeneration, confined to the middle third of the foot, especially in the outer part of the middle third. The same area is a little pale by the Weigert hæmatoxylin stain, but the degeneration is not very distinct.

Distinct recent degeneration is seen by the Marchi method within the posterior limb of one internal capsule, and is confined to a small portion of the posterior limb.

Sections from the middle part of the corpus callosum show recent degeneration by the Marchi method, but the stain has faded and is indistinct.

Recent degeneration is not seen in the right paracentral lobule.

Case VIII. (a patient in my service in the Philadelphia General Hospital).—Julia S. She was admitted to the hospital on April 30, 1903. According to the history obtained from her relatives in July, 1903, the symptoms began about three years previously, when she was forty-eight years of of age. The onset was gradual, the paralysis beginning in the right foot, and gradually the entire right lower limb became affected. At the end of one year the right upper limb also became paralyzed. During the past year she had not been able to lift her left lower limb as high as formerly. She could speak distinctly until about a year ago, and occasionally her speech was better than at other times. She had fed herself with her left hand. She had been in a wheel-chair for two years, and then confined to her bed one and a half months previous to admission. During the six months previous to her admission she had had incontinence of urine and feces. She had never complained of pain.

On July 9, 1903, the following notes were taken by Dr. T. H. Weisenburg:

The patient seems intelligent, understands questions, and talks fairly well. She has no upper teeth.

The pupils are dilated and equal, and respond to light in accommodation and, apparently, in convergence. The tongue is protruded straight, and shows no tremor and no atrophy. She shows her teeth fairly well, and there is apparently no palsy of the seventh nerve on either side.

The movements of the right upper limb are limited; she has some voluntary movement at the shoulder and elbow-joints. There are contractures present in the right upper limb, the arm being held closely to the chest, the elbow flexed almost at a right angle, the hand at a right angle to the forearm, the thumb turned inward, and the fingers extended and widely apart. Contracture at elbow can be overcome on forced movement, as well as that at the wrist. A distinct atrophy is present in the thenar and hypothenar eminences of the right hand, and in the interossei muscles. The grasp of the right hand is *nil*. The movements of the left upper limb are fairly well preserved. There is no contracture present, and apparently no atrophy. The limb seems to be spastic at shoulder, elbow, and wrist-joints. The reflexes on the right side, biceps tendon, triceps tendon, and wrist reflexes are limited on account of contractures. On the left side they are prompter than normal. The grip of the left hand is very poor. No apparent disturbance of sensation is found in the upper limbs. The right arm, two inches below the elbow, measures eight inches in circumference; the left, eight and one-quarter inches in circumference. The right arm, below middle of biceps, measures eight and one-half inches in circumference; the left, nine and one-quarter inches in circumference. The right forearm and arm are atrophied.

The right lower leg at the middle portion measures ten inches; the left, nine and one-half inches.

The movements of both lower limbs are limited, and both limbs are spastic. The reflexes, patellar and Achilles tendon, are prompt on both sides, and more so on the left side. Ankle clonus is present on each side, and is more persistent on the left side. It is impossible to obtain the Babinski sign. She apparently responds to touch and pain stimuli in both lower limbs. Incontinence of urine and feces is present.

On July 23, 1903, I dictated the following notes:

The speech is indistinct and somewhat bulbar in character. The movements of the eyeballs are free in all directions. When she is lying quietly the right side of the forehead is much more wrinkled than the left side, but when she is told to look upward the left side of the forehead is wrinkled as much as the right. She moves the left upper limb at all the joints, but the movements are slow and stiff. The left upper limb is somewhat rigid, and the rigidity varies from time to time.

The movements in the lower limbs are much impaired, and equally so on the two sides. She is able to flex slightly either leg at the hip or knee, and the movement is a little greater on the right side than on the left. She is generally emaciated. The left lower limb is almost completely paralyzed. Babinski's reflex is not obtained on either side, and there is no distinct movement of the great toe on either side. The left patellar reflex is prompt. The muscles of the leg and foot are spastic. The right lower limb is spastic, and the patellar reflex is prompt. Ankle clonus is present and persistent on the left side, but is not obtained on the right side, although it was obtained by Dr. Weisenburg three days previously. The stick of a pin in any part of the limbs causes signs of discomfort.

July 18, 1903. The movements of the eyeballs from side to side, upward and downward, are good. There is very little change of expression in the face. She lies with the mouth partly open, and presents the appearance of partial paralysis of each seventh nerve. Dr. Weisenburg says he has seen her laugh, and then she draws up both corners of the mouth very well. The tongue is protruded straight and to the normal extent; it is not atrophied, and shows no fibrillary tremor.

The left upper limb is decidedly rigid, but can be fully extended by the patient, though the movements are slow and awkward and somewhat incoordinate. A pin prick anywhere causes discomfort, but she makes very little effort to avoid it, probably because of her mental condition.

Testing with feeding cup does not show hemianopsia.

July 30, 1903. During the past three weeks the patient has lain with the knees drawn up, though they could be extended voluntarily. A necropsy was obtained August 7, 1903.

The pathological diagnosis, made by Dr. John Funke, was œdema of lungs, emphysema, chronic endocarditis, hypertrophy of left ventricle, chronic interstitial nephritis, chronic cystitis, hemorrhagic gastritis, thrombosis of left lateral sinus and of the superior portion of the internal jugular vein, softening of the middle of the second and third temporal convolutions on the left side, atrophic cirrhosis of the liver, and fibroid uterus.

The result of my microscopic examination is as follows:

Lumbar Region. The left crossed pyramidal tract is much degenerated; the right is only a little pale by the Weigert hæmatoxylin method. The nerve cells of the anterior horns stained by the thionin method are not greatly altered. In some the chromophilic elements are granular and there is a considerable amount of light yellow granular material, and in some the nucleus is displaced to the periphery. There is, therefore, a pathological condition of some of the nerve cells, but it is not intense. Very slight degeneration by the Marchi method is found in each crossed pyramidal tract.

Cervical Region. Here also the left crossed pyramidal tract is much degenerated, although not completely so, while the right crossed pyramidal tract is scarcely degenerated at all. The direct pyramidal tracts are not degenerated. The degeneration in each crossed pyramidal tract by the Marchi method is very slight, showing that the process was a chronic one. The cells of the anterior horns by the thionin stain are like those of the lumbar region, except in the right anterior horn, where the alteration is more intense and considerably more so than in the left anterior horn. No meningitis is found anywhere.

Medulla Oblongata. The right anterior pyramid is slightly degenerated, as shown by the Weigert hæmatoxylin method.

Pons. The pyramidal tract fibres on each side are well stained, but some of those in the right side are a trifle less deeply stained.

Cerebral Peduncles. A small area less deeply stained is found in the middle of the foot of the right cerebral peduncle. It is only in the posterior part of the foot, and does not extend more than half way between the periphery and the substantia nigra.

The clinical picture presented by Case VIII. was a striking one. According to the history of the patient, paralysis of the limbs had begun about three years before her death. Relatives of the patient stated that when the woman was forty-eight years old weakness began in the right foot and gradually the entire right lower limb became affected, and that at the end of one year the right upper limb also became paralyzed. During the year previous to her admission to the hospital she became unable to lift the left lower limb as well as she had done formerly.

When she came under my observation, in July, 1903, both lower limbs were spastic and paralyzed, but the voluntary movement was a little less in the left lower limb. Ankle clonus was persistent on the left side, but was not obtained by me on the right side. The tendon reflexes were exaggerated in both lower limbs, but more so in the left limb. The whole body was emaciated. The right upper limb was almost completely paralyzed and much contractured, the arm being held closely to the chest, the forearm flexed on the arm almost at a right angle, the hand flexed at a right angle to the forearm, the thumb turned inward, and the finger extended. There was some voluntary movement at the right shoulder and elbow, but the grasp of the right hand was *nil.* The thenar and hypothenar eminences and interossei muscles of the right hand were atrophied. The movements of the left upper limb were fairly well preserved, and the limb was neither contractured nor atrophied, although it was spastic, and the tendon reflexes were exaggerated. Sensation for touch and pain was normal in all the limbs. The woman had incontinence of urine and feces. Speech was indistinct and somewhat bulbar in character, and was said to have been so about a year. The cranial nerves otherwise did not appear to be affected.

This was, therefore, almost a case of triplegia, as the left upper limb was comparatively little affected, but a triplegia in which the right upper limb was most implicated, and the left lower limb was more affected than the right lower limb, as shown by the greater exaggeration of the tendon reflexes and the slightly greater loss of voluntary power in the left lower limb.

The microscopic findings were at first somewhat of a surprise. As the paralysis was said to have existed longer in the limbs of the right side I expected to find the right crossed pyramidal tract more degenerated, but it was very slightly diseased, while the left was much degenerated, but by no means completely so. In these findings we have an explanation for the greater implication of the left lower limb as compared with the right, but it was not until the thionin stain was employed that a satisfactory explanation of the intense paralysis of the right upper limb could be obtained. By this stain the nerve cells of the right anterior horn of the cervical region were found intensely degenerated, and much more so than those of the left anterior horn.

This case shows that great spasticity of the limbs may be caused by degeneration of the pyramidal tracts that is barely detectable, as the right crossed pyramidal tract in this case was only a little degenerated, and yet the spasticity of the right lower limb was great. This is very interesting and instructive in connection with the famous case reported by Senator[1] and later by Wolff,[2] in which spasticity seemed to be present without degeneration of the central motor tracts.

The case further shows that where a limb is spastic from slight degeneration of the crossed pyramidal tract, as the right upper limb of my patient, disease of the cells of the anterior horn on the same side may assist in the formation of contracture by increasing the paralysis. Where the nerve cells of the anterior horns are diseased without degeneration of the central motor tracts, as in anterior poliomyelitis, the paralysis is flaccid, and contracture, if it occurs, is usually from faulty position or overaction of less affected muscles. If, however, the central motor tracts are diseased, Case VIII. would seem to indicate that degeneration of the nerve cells of the anterior horns may increase the tendency to contracture.

Case VIII. further shows, provided we accept the history given by the relatives of the patient, that we cannot always judge of the longer duration of degeneration in any tract by the degree of the degeneration, because in this case the right crossed pyramidal tract was much less diseased than the left, and yet the paralysis is said to have begun on the right side. The very slight recent degeneration of the right crossed pyramidal tract, as shown by the Marchi method, indicates that the degeneration was of long duration. We might assume that the degeneration of the left crossed pyramidal tract existed for some time before it caused distinct symptoms, and was more progressive than that of the right side.

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[1] Deutsche med. Wochenschrift, 1894, vol. xx. p. 443.

[2] Zeitschrift f. klin. Med., xxv. p. 326

The absence of marked degeneration by the Marchi method in the central motor tracts in Case VIII., shows that in some cases of amyotrophic lateral sclerosis the degeneration may be of relatively slow development, and it is exceedingly desirable to ascertain why in some cases the degeneration is slow and in others cases very rapid. The solution of this problem I have not been able to find, but my cases show that when degeneration of the nerve cells is associated with degeneration of the motor fibres the course of the disease is usually more rapid, and this is especially true if bulbar symptoms develop early.

Most of the cases (Cases I., III., VI., VII., and VIII.) show that disturbance of the bladder and rectum may be caused by degeneration of the central motor tracts, even where the posterior columns and the nerve cells of the anterior horns are not diseased, and that degeneration of the posterior columns is not to be diagnosed simply because this sign exists.

I do not believe that amyotrophic lateral sclerosis is a toxic process, but that it should be regarded as an abiotrophy. I think it is doubtful that the process in most cases is ascending in each nerve fibre attacked, and the cessation of the degeneration in the medulla oblongata or pons is no proof of an ascending process. No one, so far as I know, has described a case of amyotrophic lateral sclerosis with necropsy in which the cervical region was normal, and degeneration was found in the pyramidal tracts below this region. It seems to me reasonable to believe that there is a gradual death of the portions of the central motor neurones most remote from the cells of origin, and that the extent of the nerve fibres diseased varies in different cases, but that the whole portion of each fibre that is affected is diseased usually almost simultaneously, although the individual nerve fibres become affected at different periods. So far as I know Hoche[1] alone has found that the Marchi method did not stain to any extent the pyramidal tracts of the cord, medulla oblongata, and pons in a case of amyotrophic lateral sclerosis, but showed numerous black dots in the crusta. It is doubtful, however, whether those fibres that showed long-standing degeneration in the lower part of the pyramidal tract were the same that showed recent degeneration higher up.

I had the opportunity two or three years ago to observe the death of a tree in front of my house. In the spring no leaves appeared on the branches, except on the portions near the trunk, although during the previous fall the tree had appeared to be healthy, so that the peculiar appearance was presented of a tree dead in the terminal branches, but alive in its trunk and in the branches near the trunk; that is, in the portion nearest to the root from which nourishment was received. Something very like this occurs in amyotrophic lateral sclerosis.

In some respects amaurotic family idiocy resembles amyotrophic lateral sclerosis, and it also is I[1] believe an abiotrophic process. The nerve cells and nerve fibres degenerate early, and in the case that I have recently studied by microscopic sections the Marchi method showed more intense recent degeneration in the foot of the cerebral peduncle than in the motor tracts of the spinal cord. These are like the findings of Hoche in amyotrophic lateral sclerosis, but it is not necessary to assume that in the same nerve fibres degeneration occurred in different portions of the fibres at different periods.

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[1] Archiv für Psychiatrie, xxviii. p. 980.

Case VIII. differs in some respects from Case IV., the case reported in 1903 by Dr. Mills and myself. In Case VIII. the right thenar and hypothenar eminences and the right forearm were atrophied, but the corresponding parts of the left upper limb were not atrophied. In Case IV. muscular atrophy was not present. In both cases the limbs of the right side were first affected, and in both the left lower limb later became paralyzed. Both cases presented the clinical signs of progressive degeneration of the central motor tracts, beginning in each on the right side of the spinal cord and later involving the left side, and beginning especially in the nerve fibres affording motor innervation to the right lower limb.

In Case IV. the degeneration of the cross pyramidal tract was more intense on the right side of the cord, whereas in Case VIII. it was more intense on the left side of the cord, and the greater paralysis of the right upper limb finds an explanation in the more intense degeneration of the nerve cells of the anterior horn on the right side. The lower limbs were nearly equally affected. Case IV. belongs to the type of primary lateral sclerosis,

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[1] American Journal of the Medical Sciences, January, 1905.

Case VIII. to the type of primary lateral sclerosis plus degeneration of the nerve cells of the anterior horns, viz., amyotrophic lateral sclerosis.

In the case reported by Patrick[1] the unilateral paralysis began when the patient, a girl, was about fourteen years of age, and seems to have been observed in the right upper and lower limbs simultaneously. Patrick says: "The apparent flaccidity, as shown by the dangling arm, foot-drop, and moderate steppage, and the presence of real spasticity, as shown by great exaggeration of the reflexes and Babinski's sign, present a very peculiar clinical picture. The case is entirely unique in my experience, and an inconsiderable search of the literature has disclosed nothing just like it. Progressive muscular atrophy, amyotrophic lateral sclerosis, syringomyelia, multiple sclerosis, muscular dystrophy, neuritis, spinal tumor, and hysteria may easily be excluded. It is difficult to imagine any gross cerebral lesion which could cause all the symptoms of this case and yet cause no sensory disturbance, no headache, vomiting, dizziness, or optic neuritis, no more involvement of cranial nerves, no spasm, twitching, tremor, or inco-ordination, no interference with speech, and no mental deterioration. Degeneration of the upper motor neurones would seem best to explain the condition, but there is not nearly so much spasticity as in primary lateral sclerosis, and there is more diminution in muscular volume than I have seen in cases of that disease. I am unwilling to make a diagnosis. Perhaps I would better say I am unable, but I think the case more closely conforms to the clinical type proposed by Dr. Charles K. Mills than to any form of disease with which I am acquainted, and his hypothesis of the pathology, 'a slowly increasing degeneration of the pyramidal fasciculi or of the cerebral motor neurone system,' seems to suit the symptoms of my patient."

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[1] Journal of Nervous and Mental Disease, August, 1903, p. 469.

The paralysis in Patrick's case was not ascending, and in this respect the case differed from the type of unilateral progressive ascending paralysis described by Dr. Mills, although I do not think it should be excluded on this account. The case was not with necropsy.

Patrick refers to a case of unilateral progressive ascending paralysis reported by me, but does not regard my case as conforming to the type because optic atrophy was present on the affected side and the patient occasionally found some difficulty in retaining the feces. In most of the cases described in my present paper disturbance of bladder or rectum was present, so I cannot understand why this sign might not occur in an unilateral degeneration of the pyramidal tract. Optic atrophy does not belong to this type, but is not, in my judgment, sufficient to exclude the case. Optic atrophy may occur without any other sign of disease of the nervous system, and in my case was unilateral and slight. Therefore, while I have very great respect for Dr. Patrick's opinion, I see no reason why the clinical case described by me in 1901 should not be classed with those of unilateral progressive ascending paralysis, probably resulting from primary degeneration of one pyramidal tract. Where should we class the following case, in which optic atrophy was intense, if not among the cases of primary degeneration of the pyramidal tracts? It hardly appears like a case of multiple sclerosis.

The patient, a soldier, aged thirty-seven years, was referred to me by Dr. E. B. Block, of Atlanta, Ga., and Dr. H. M. Thomas, of Baltimore, Md. He denies syphilis, and his wife has given birth to a healthy child now fourteen months old. There are no hereditary diseases in the family. In the spring of 1903, about four to six weeks after he had injured his right foot slightly, he noticed that the muscles of the right leg, and especially those of the thigh, would tremble when he was standing at attention. The right lower limb became gradually weaker after June, 1903, and at this period the left lower limb was not affected. About August, 1903, he began to have what he called "cramps" in the right lower limb. In November or December, 1903, he found that his right hand was not so powerful as the left. He never had any sensory disturbances in any of the limbs. He does not pass urine so freely as formerly. Sight began to fail in 1901, but it was not until April, 1904, that he noticed any distinct and persistent disturbance of vision. He has never had diplopia, vertigo, nausea, vomiting, nor severe headache.

The cranial nerves, with the exception of the optic, seem to be normal. The grasp of the right hand is not so strong as that of the left, and he cannot move the fingers of the right hand so freely as those of the left hand. The biceps and triceps tendon reflexes are prompter on the right side.

The limbs are not wasted. The patellar reflex on each side is much exaggerated, but more so on the right side, and patellar clonus is obtained only on the right side. There is almost ankle clonus on the right side, and the Achilles tendon reflex is a little exaggerated on the left side. The right lower limb is very spastic, the left is not. Babinski's reflex is distinct on each side, but more so on the right side. Resistance to passive movement is diminished in the right lower limb, but is about normal in the left lower limb. Gait is spastic in the right lower limb, and this limb is swung in walking. No ataxia is observed. No objective nor subjective disturbances of sensation are present.

Dr. Alexander Duane, of New York, made a very full report on the ocular condition July 28, 1904, in which he says: "His R. V.=at first $\frac{15}{200}$, later on steady looking rose to $\frac{15}{60}$. L. V.= movements of hands at 1 $(=\frac{1}{1000})$. . . . Disks uniformly pale, somewhat opaque looking, not shrunken. Vessels unchanged, except that arteries are rather small. There are no evidences of preceding neuritis or retinitis, and the appearances to my mind are decidedly those of a primary optic atrophy. In the left eye the temporal side of the disk is slightly paler than the nasal, but not more so than one would find physiologically. The appearances, in fact, are those of a uniform atrophy." Dr. Duane thought that the optic atrophy was like that of tabes.

No cases except those mentioned in the paper by Dr. Mills and myself, that could be regarded as belonging to the type of primary uncomplicated unilateral degeneration of the pyramidal tract, have been reported so far as I know. One of the cases which I now put on record (Case VIII.) shows that what is true of the bilateral form of primary degeneration of the pyramidal tract is true also of the unilateral form, viz., that the nerve cells of the anterior horns of the spinal cord are not likely to escape degeneration. Last year Dr. Mills and I were able to show by a microscopic study of the brain and spinal cord that an uncomplicated primary degeneration of the pyramidal tract of one side may occur, and without degeneration of the nerve cells of the anterior horns, and this case, so far as I know, is unique, but even in this case the degeneration was not long confined to one side of the cord. Now, I am able to demonstrate that the same clinical signs plus atrophy may be caused by a primary degeneration of the pyramidal tract of one side plus degeneration of the nerve cells of the anterior horns.

I have endeavored to find cases of this unilateral form of amyotrophic lateral sclerosis reported in the literature, and have found ten, but not all with necropsy. It is not improbable that some other cases may be found, but the unilateral form of amyotrophic lateral sclerosis is not common, and always bilateral implication of the spinal cord occurs within a short time. In these ten cases only three were of the ascending type, the weakness commencing in one lower limb and later extending to the upper limb of the same side. These were the cases of Vierordt, Mott, and Probst. The ten cases are as follows:

In Dejerine's case (Case II. of his paper)[1] labioglossolaryngeal paralysis occurred in a woman, aged seventy-four years, and was associated with slight right hemiplegia. The bulbar symptoms were first noticed in May, 1878. In October, 1879, there was no localized atrophy, but the whole body was much emaciated, and equally so on the two sides. The right lower limb was weak, but not contractured, and the woman was obliged to use her hands in order to cross her right lower limb over the left. The left lower limb was not weak. The right upper limb was a little feeble. The slight right-sided paresis of the limbs was, therefore, more intense in the lower limb. The woman died about three weeks after this examination was made. The drawings of the cervical region of the spinal cord show that the pyramidal tracts were symmetrically degenerated.

In a case reported by A. Pick[2] weakness of the left upper limb began about April, 1875, and was soon followed by weakness of the left lower limb. About nine months after the left upper limb became weak the right limbs became paretic. The crossed pyramidal tracts were much degenerated.

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[1] Archives de Physiologie, 1883, vol. ii. p. 200.

[2] Archiv für Psychiatrie, vol. viii. p. 283.

In Vierordt's[1] case weakness of the right lower limb began in August, 1878, and in the beginning of 1879 weakness of the right upper limb was noticed. Atrophy of the right thenar and interossei muscles then developed, and in 1880 the left upper and lower limbs became paretic. The crossed pyramidal tracts were much degenerated.

In the fourth case of Leyden, as quoted by Probst,[2] the patient was a woman, aged fifty-three years. Bulbar symptoms developed first. A year later weakness of the left upper limb occurred, and this was soon followed by weakness of the left lower limb. The duration of the disease was two years.

In Lennmalm's[3] case bulbar symptoms occurred first in the fall of 1885. In the spring of 1886 the right limbs became weak and paresis increased. In the summer of 1886 the left upper limb began to get weak, and muscular atrophy developed. The case was with necropsy.

In Mott's[4] case the duration of the disease was one year. The commencement was with weakness and numbness in the right leg, gradually increasing, and attended with wasting of muscles and exaggerated deep reflexes followed by similar affection of right arm and hand, with especial wasting of thenar, hypothenar eminences and interossei muscles. A little later the left leg became affected in a similar manner to the right, and then the left arm and trunk. The termination was with bulbar symptoms and probably paresis of the diaphragm. The sclerosis of the pyramidal tracts in this case seems to have been symmetrical, as shown by the photographs.

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[1] Archiv für Psychiatrie, vol. xiv. p. 391.

[2] Ibid., vol. xxx. p. 766.

[3] Abstract in the Neurologisches Centralblatt, 1887, p 550.

[4] Brain, 1805, p. 21.

In the case of Senator, as cited by Probst, the patient, a woman aged fifty-seven years, began to grow weak in the left limbs, later the right limbs became affected, and bulbar symptoms developed. The duration of the disease was five years.

In Probst's[1] third case bulbar symptoms appeared first. The right upper and lower limbs became weak. At the time the case was reported it seems that the left limbs were not very weak. Muscular atrophy developed. A necropsy was not obtained.

In his fifth case, also without necropsy, the symptoms began with weakness in the left upper and lower limbs, and later the right upper limb became weak. Muscular atrophy developed.

In his seventh case, without necropsy, weakness of the left lower limb developed. After a few months the left upper limb also became weak. Bulbar symptoms and muscular atrophy developed. A few months later the right upper limb became weak.

In his eleventh case, without necropsy, the first symptoms were bulbar. Soon the left upper limb became weak, and within a short time the left lower limb also became weak. Atrophy developed. Later the right side became affected, but the history is not very complete.

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[1] Archiv für Psychiatrie, vol. xxx.

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A PATHOLOGICAL STUDY OF AMAUROTIC FAMILY IDIOCY.[1]

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Amaurotic family idiocy is a disease that has been studied especially by American physicians. Falkenheim[2] after a careful examination of the literature has found 64 cases reported (1901), of these 47 were seen by physicians, 30 cases (24+6) occurred in America, 11 (9+2) in England, and 23 (14+9) on the Continent of Europe.

More recently (1904) Heveroch[3] has collected the records of 86 cases, of which 44 occurred in America.

B. Sachs was the first to recognize the clinical symptoms of amaurotic family idiocy, and gave the disease the name by which it is now known. It was subsequently shown that one symptom of the condition had been previously recognized by Warren Tay, who described a case presenting "symmetrical changes in the region of the yellow spot in each eye of an infant."

In his earlier publication on this subject in 1887, Sachs supposed the disease to be due to peculiar changes in the cortical cells. Hirsch was the first to show that not only the cortical cells, but all the cells of the gray matter of the entire central nervous system were similarly affected.

I have had the opportunity to make a pathological study in a typical case of the disease that had been under the care of Dr. J. H. McKee, and had also been examined by me and others. I am much indebted to Dr. McKee for the pathological material. His necropsy notes are as follows:

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[1] Read in abstract at the meeting of the American Association of Pathologists and Bacteriologists, April, 1904.

[2] Jahrbuch f. Kinderheilkunde, 1901, p. 123.

[3] Abstract in Neurologisches Centralblatt, October 16, 1904, p. 948.

"A piece of nerve was cut from the internal plantar; a piece of muscle from the flexor brevis digitorum.

"Thymus was small.

"Thyroid was well developed and presented a large isthmus.

"Pericardium: There was slight increase in the amount of the fluid.

"Heart: Both post-mortem and chicken-fat clots were found, but there were no valvular or endocardial lesions.

"Lungs: The left lung showed a massive catarrhal pneumonia of the lower lobe, and of the extreme posterior portion of the upper. The apex and free anterior border exhibited considerable emphysema. The basal portion of the lower lobe sunk in water. The right lung showed similar pneumonic changes in the lower and middle lobes.

"Pleuræ were free from adhesions or recent exudate.

"Peritoneum: Nothing abnormal was noticed, and there was no undue dryness.

"Appendix was very long and thin.

"Intestine: The small bowel exhibited intussusception (agonal?) at five places. The large bowel was small and collapsed.

"Spleen was slightly enlarged but not unduly dark. It presented no notch.

"Liver also was slightly enlarged, but remarkable in the fact that it presented none of the naked eye appearances of fatty infiltration.

"Kidneys: Nothing of interest was noticed macroscopically. Suprarenals were apparently normal.

"Lymph nodes: The peribronchial glands were enlarged and red, but the process was obviously an acute one. No caseous lymph nodes were noticed either in the thorax or abdomen.

"Brain: The subarachnoid fluid was much increased in amount. The meningeal vessels were intensely injected. The brain itself was very moist and heavy."

The results of my microscopic study are as follows:

The nerve cells throughout the spinal cord are much enlarged, the chromophilic elements have disappeared except about the nucleus, and here they have become granular, and in many cells the nucleus has been displaced to the periphery of the cell body. The dendritic processes of the nerve cells have disappeared in most of the cells.

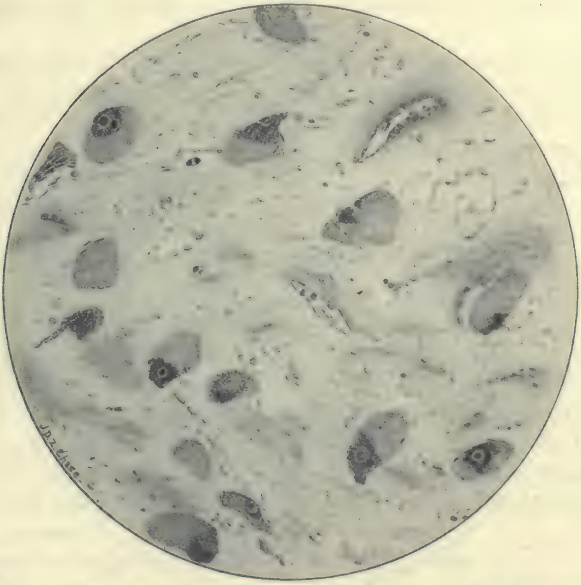
The cells of the hypoglossus nucleus, of the nucleus ambiguus, of the lower olive, of the abducens nucleus, of the trigeminus and oculomotorius nuclei are in a similar condition.

The cells of Purkinje in the cerebellum are intensely altered, enlarged, and with the exception of the nucleus are structureless, so that they are found with difficulty.

The nerve cells of the paracentral lobule, the small cells as well as the cells of Betz, and those of the frontal and occipital lobes are in a similar condition.

I have not been able to find a single normal nerve cell in the central nervous system.

FIG. 1.



Cells of the anterior horn in the lumbar region in a case presenting the symptoms of meningitis. The pathological changes consist of an alteration of the nerve cells like that of amaurotic family idiocy. The cells are represented as they appear in one field of the microscope, in order to show the intense alteration of all the cells.

The Marchi method shows most intense alteration of the white matter in the brain and cord, especially in the former.

The crossed pyramidal tracts do not contain black dots in the lumbar region, as the degeneration here is of long standing. The nerve fibres of the anterior horns show some degeneration by the Marchi method.

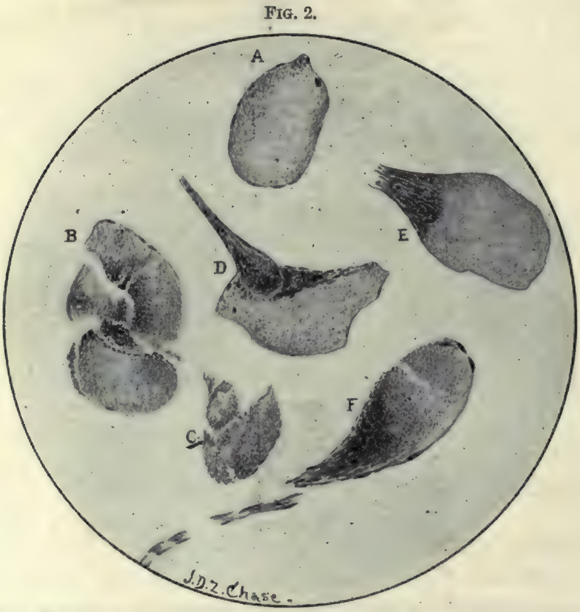
The crossed pyramidal tracts in the midthoracic region show a little recent degeneration, as do also the direct cerebellar tracts.

The same condition is found in the lower cervical region. The posterior columns are not degenerated.

The nerve fibres within the lower olives are much degenerated.

In the pons the intramedullary portion of the trigeminus, the lemniscus, the fibres crossing in the raphe, the superior cerebellar peduncle are much degenerated. The degeneration of the motor tracts is of longer duration than is that of many of the other tracts of the pons, but is still very intense, and much more so than is the degeneration of the pyramidal tracts in the cord as shown by the Marchi method. Indeed, an examination of the sections leads one to conclude that the motor tracts may have degenerated earlier in the cord than in the brain.

FIG. 2.



Cells from the same case as represented in Fig. 1, selected from the anterior horns of the lumbar region, with the exception of cell *E*, which is from the paracentral lobule. Cell *A* shows complete chromatolysis. Cells *B* and *C* are broken into several fragments. In cells *D*, *E*, and *F* the chromophilic elements are seen only in one part of the cell body, and the nucleus is not distinct. By comparing Figs. 1 and 2 with the illustrations in the paper by Hirsch and Sachs it will be seen that the alterations are exactly like those of amaurotic family idiocy.

The foot of the cerebral peduncle is much degenerated, but the degeneration is of long duration, and is more intense in the dorsal portion of the foot, and here the fatty granular cells are very numerous, especially about the bloodvessels. There is comparatively little free degenerated myelin in the foot of the peduncle, and the numerous fatty granular cells stain deeply by the Marchi method.

This seems to indicate that the degeneration had existed long enough for the myelin to be absorbed by these cells.

Intramedullary fibres of the oculomotor nerve show a moderate amount of recent degeneration by the Marchi method.

The superior cerebellar peduncle is much degenerated and contains much free myelin.

The posterior longitudinal bundle may be slightly degenerated.

The lemniscus is considerably degenerated.

The white matter of the occipital and frontal lobes and of the paracentral lobule is much degenerated by the Marchi method.

Sections taken from the middle of the upper part of the corpus callosum are intensely degenerated by the Marchi method.

A piece of muscle from the foot, the flexor brevis digitorum, stained by the Marchi method shows no recent degeneration, but a piece from the same muscle stained by eosin and hæmalum shows an increase in the number of connective-tissue nuclei and probably also in the number of the sarcolemma nuclei. The muscle fibres appear degenerated, although I have no normal muscles from a child of about the same age for comparison.

A piece of one internal plantar nerve stained by the Marchi method shows considerable degeneration, but a piece from the same nerve stained by the Weigert hæmatoxylin method or acid fuchsin is not degenerated or at least very slightly degenerated.

Sections stained by the Weigert hæmatoxylin method or acid fuchsin also show degeneration of the central motor tracts. The anterior and posterior roots of the cervical and lumbar regions are not degenerated.

No signs of inflammation are found anywhere.

Falkenheim found that necropsies had been obtained in 11 cases: in 2, however, no report was made—those of Grosz. The 9 cases with reports of necropsies were those of Knapp and Sachs, Kingdon (3), Peterson, Hirsch, Mohr, and Clairborne. The report of necropsy in Clairborne's case is exceedingly brief. Since Falkenheim's paper appeared cases with necropsy have been reported by Frey (1), Schaffer (1), and Sachs (1). In Mohr's case the report of the histological examination was written by Schaffer.

Falkenheim says that in amaurotic family idiocy the degeneration of the pyramidal tracts has been followed through the cerebral peduncles, pons, medulla oblongata, and spinal cord. The lemniscus also has been found degenerated, as well as the tangential fibres, the spinal root of the fifth nerve, the anterior cerebellar peduncle, the columns of Goll, and the optic nerves. Falkenheim mentions

Hirsch's paper, but he does not do justice to the latter's cellular findings.

Schaffer[1] found intense and widely diffused degeneration of the brain and degeneration of the pyramidal tracts of the spinal cord. The gray matter of the cord was not diseased. No mention is made of the condition of the nerve cells of the brain. This is not the same case as the one described in Mohr's paper.

Frey's case is especially interesting because his is one of the very few in which the Marchi method has been employed in the study of a case of amaurotic family idiocy. He found increased consistency of the brain and spinal cord. The crossed pyramidal tracts, the tracts of Gowers, and the lateral border zones (Grenzschichten) were degenerated. The direct cerebellar tracts and the anterior columns were normal. The degeneration of the posterior columns, especially of the columns of Goll, began in the lower thoracic region and could be followed to the nuclei of the columns of Goll. The fibre reticulum of the anterior horns had almost disappeared in the cervical region. The white matter of the brain showed much degeneration. It is probable that the peculiar cellular alterations of amaurotic family idiocy were present in this case, although Nissl's method could not be employed. Frey[2] says that the pyramidal cells of the cortex were rounded and had lost their normal shape. There was no trace of inflammation. Evidently the peculiar cellular alteration of this disease escaped him, as he does not speak explicitly about it, and does not appear to have read Hirsch's paper. The optic tract and chiasm were degenerated but the optic nerves were normal. The entire central nervous system was affected.

He refers to another case of the disease that he was studying in which a pronounced degeneration of the nerve cells with vacuolation was found, but there seems to have been little to him that was characteristic in this cellular alteration.

Sachs'[3] recent paper fully confirms the extraordinary cellular findings described by Hirsch in a case of amaurotic family idiocy, and these seem to be the only cases in which this peculiar and widespread cellular degeneration was reported, except that Sachs now believes that some of his earlier cases showed a similar alteration.

The description of the changes of the nerve cells in my report conforms very closely with those given by Hirsch[4] and Sachs.

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[1] Wiener klin. Rundschau, 1902, p. 324.

[2] Neurologisches Centralblatt, September 16, 1901, p. 836.

[3] Journal of Nervous and Mental Disease, 1903, p. 1.

[4] Ibid., 1898, p. 538.

If the widespread and intense cellular alteration is a common condition in amaurotic family idiocy it is remarkable that, according to Falkenheim, the electric irritability of the muscle is preserved, and that only Koplik and Grosz have found slowing or diminution of the faradic contractility.

Russell and Kingdon[1] stated in 1899 that no changes in the peripheral nerves had been found in amaurotic family idiocy, and I have not been able to find the report of any such changes recorded in the literature.

In the case that I have studied the piece removed from the internal plantar nerve was distinctly degenerated by the Marchi method.

I do not share Sachs' opinion that the changes in the nerve cells in the cases of Rolly, Collier, and Mya and Levi were like those of amaurotic family idiocy. Rolly[2] says that in the lumbar region the processes of the ganglion cells were abnormally intensely stained, and the portion of the processes that normally does not stain took the stain. The nerve cells were least altered in the lumbar region. Near ganglion cells that were entirely normal were others in which chromatolysis had occurred, in some this was perinuclear while the chromophilic elements at the periphery of the cell were preserved. Some cells showed peripheral chromatolysis while the chromophilic elements about the nucleus were preserved. Nowhere was there any marked alteration of the form of the cell.

The changes in the nerve cells of the thoracic region were like those of the lumbar region.

The changes in the cervical region were more intense. Normal cells were less numerous than elsewhere in the spinal cord, and chromatolysis was more pronounced. Still more intense were the changes in the brain. Nowhere does Rolly speak of a swelling of the cell, with destruction of the cell processes, and complete loss of all stainable portions of the cell except about the nucleus, which is displaced to the periphery. Nor was the alteration by any means found in all the nerve cells. The alteration of the nerve cells was pronounced in Rolly's case, but, as a study of his drawings show, it was very unlike that of amaurotic family idiocy. The pyramidal tracts were perfectly normal.

In two cases, one of cerebral diplegia and one of generalized rigidity, with necropsy reported by J. S. Collier,[3] the cellular alteration was not like that of amaurotic family idiocy. The pathological report in these two cases is by Risien Russell. In the first case there was a great paucity of nerve cells in the cerebral cortex. Neither the cells nor their nuclei presented any signs of swelling. Many of the cells preserved more or less their normal pyramidal shape, but they were all below the normal in size, many of them very much so. The cells stained diffusely by the Nissl method. The pyramidal tracts were degenerated.

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[1] Allbutt's System of Medicine, The Macmillan Co., 1899, vol. viii. p. 728.

[2] Deutsche Zeitschrift f. Nervenheilkunde, Bd. xx. p. 152.

[3] Brain, 1899, vol. xxii. p. 378.

In the second case the cellular alteration and the degeneration of the pyramidal tracts were very similar. The changes in the nerve cells in these cases were not at all like those of amaurotic family idiocy.

So far as I can determine the changes in the nerve cells described by Mya and Levi were not those of amaurotic family idiocy.

In 1899, in association with Dr. D. J. McCarthy,[1] I reported a case not of amaurotic family idiocy, with changes in the nerve cells, however, like those occurring in this disease. The patient, a girl, was nine years old at the time of her death. Her sight was said to be good, but it is not known that any visual examination was made. She could not speak. She had a normal gait earlier in life. She began to walk before she was two years of age and had no motor paralysis. She never had convulsions. The feet and hands were always cold. She was said to be lacking in sensation, and would handle hot coals or hot food without making a sound of discomfort. She was exceedingly active and noisy, wet her bed and day clothing, did not understand what was said to her, and could not read. During the year previous to her death she showed gradual emaciation, but no cause could be detected for this. The gait was observed to be more shambling than previously. Emaciation became extreme before death.

At the necropsy the bridge of the nose was found flattened, the lower front teeth were nicked like those known as Hutchinson's teeth, the sternum was prominent. Internal hemorrhagic pachymeningitis was found on each side of the brain. The pia was very œdematous and the brain was quite soft. The nerve cells of the paracentral lobule, of the frontal lobe and posterior portion of the parietal lobe, of the cerebellum and bulbar nuclei, and of the anterior horns of the cervical and lumbar regions were swollen and altered in the manner described more in detail in the following case. The capillaries in the cerebral cortex and in the spinal cord were very numerous and congested. Secondary degeneration in the spinal cord was not present, even by the Marchi method.

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[1] **Journal of Nervous and Mental Disease, November, 1899, p. 677. Spiller, ibid., 1901, p. 140.**

These remarkable cellular changes throughout the central nervous system were described in detail and were represented by drawings. Reference also was made in that paper to the similar condition described by Hirsch in amaurotic family idiocy. Microscopic examination showed that in our case the optic nerves were normal. It was thought that the œdema of the cortex possibly might explain the alteration of the nerve cells, although it was acknowledged that the cases of uræmia reported in literature did not justify this view.

In 1900 I observed again changes in the nerve cells that were exactly like those occurring in amaurotic family idiocy in another case that could not be considered as typical of this disease. The patient, a boy, was within three months of eight years of age at the time of his death. The notes were incomplete, but it is recorded that he was of feeble mental development, did not know the alphabet, did not talk, and could not dress himself properly. His sight and hearing were said to be good, although it is doubtful whether there was a visual examination. His gait was spastic. He was small and had scoliosis. Sensation was normal so far as could be determined. He did not have convulsions. He died with symptoms of cerebrospinal meningitis.

At the necropsy a large amount of cerebrospinal fluid escaped when the calvarium was removed. The brain was œdematous. The microscopic examination of the nervous tissues showed little evidence of inflammation. The nerve cells throughout the central nervous system were greatly altered, those in the posterior as well as those in the anterior horns of the spinal cord, the cells of the nuclei of the cranial nerves, sensory as well as motor, the cells of Purkinje and those in the parietal lobe, especially the cells of Betz, were all affected. The cell-body everywhere was swollen and rounded, many of the dendritic processes had disappeared; the chromophilic elements were seen only in one portion of the cell, and here they surrounded the nucleus, and even here they were more or less disintegrated, and the nucleus was at the periphery of the cell-body. Numerous small bacilli were found within the nervous tissues, but these may have entered the tissues after death.

In neither of these two extraordinary cases was secondary degeneration found, even by the Marchi method. Both children were of very feeble mentality, and were from the Pennsylvania Training School for Feeble-minded Children. It is incomprehensible that any function could be possible with such intense and general alteration of the nerve cells, and it is improbable that such an alteration could have existed a long time.

No one has been able to make a thorough microscopic study of the early degeneration of amaurotic family idiocy, and in those cases in which a microscopic examination has been made the disease had reached its full development and had been the cause of death. What the primary alterations are in this disease we do not know, but the widespread cellular alteration and comparatively slight changes in the white matter in the two cases referred to by me suggest that the cells probably are first diseased in amaurotic family idiocy, but this is by no means certain.

I cannot, any more than does Sachs, accept the view that amaurotic family idiocy is a toxic or acquired disease. Sachs, in referring to my second paper, remarks that I seem to interpret the cell changes as the result of the acute process in the cases to which I have referred above. The sentence to which he refers probably is this: "A theory of a toxic condition in the second case could be well supported by the clinical history, but not so well in the case of internal hemorrhagic pachymeningitis, although even in this it could not be absolutely rejected." I think I imply a very decided doubt regarding the nature of the process. Indeed, I feel unable to say why in the two cases to which I have referred such an extraordinary pathological condition was found. These two cases are unique in the literature so far as I know, and, therefore, their interpretation at present should be left in doubt. They show a remarkable resemblance in their pathological findings to those of amaurotic family idiocy, and I am tempted to suggest that *possibly* they were aberrant types of this disease. In neither case was the clinical history complete and in neither case was an examination of the eye-grounds made.

It is important in this connection to mention that the first child to whom I have referred, who was the third child in the family, had a sister three years her senior, the second child in the family, who also died in a home for feeble-minded children, at the age of eight years, from what was supposed to be "tuberculous meningitis." It is very doubtful whether a microscopic examination or even a careful gross examination was made in this case. The two sisters were said to be so alike that the parents had only one photograph taken as representing both children. In my second case death occurred following symptoms suggestive of meningitis, and a number of children with amaurotic family idiocy have died with symptoms of meningitis.

Can there be a type of disease resembling closely amaurotic family idiocy but causing death about the eighth or ninth year of life, instead of within the second year of life? It seems to me that this is possible, because the alteration of the cells throughout the nervous system, in the two cases I have referred to, is entirely unlike anything occurring in other forms of imbecility and spasticity, and is so striking that anyone who has seen it once can never mistake it.

Russell and Kingdon say that two cases of amaurotic family idiocy have been reported in which death did not occur in the earliest years (they are those of Koller[1] and Peterson[2]), but Falkenheim remarks that these two cases were the same, so that really there was only one with a delayed fatal termination. The case is not with necropsy. Peterson mentions that in this case blindness was noticed when the child was six months old. At the time Peterson wrote his paper the child was five and a half years old and was an idiot. Falkenheim says that in 64 cases of amaurotic idiocy reported, death is mentioned as occurring in 38. Two patients died in the first half of the third year of life, one when three and a half years old, and the others before the end of the second year.

It is doubtful whether the case reported by Hirschberg[3] should be regarded as one of amaurotic family idiocy. His patient was fifteen years of age.

Falkenheim mentions that the characteristic ocular changes were not found in all cases of amaurotic family idiocy, so that the uncertainty concerning the existence of ocular changes in my two cases is of less importance. The absence of ocular changes must make the diagnosis difficult.

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[1] New York Medical Record, 1896, p. 266.

[2] Journal of Nervous and Mental Disease, 1898, p. 529.

[3] Centralblatt für Augenheilkunde, January, 1904.

THE PATHOLOGY OF CEREBELLAR TUMORS.*

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It is not the purpose of this paper to consider minutely the histology of cerebellar growths, as this information can be obtained in any textbook on neurology. The pathological aspects of the various conditions which give the symptoms of cerebellar tumor will be considered, especially in a surgical sense.

It is difficult to make a satisfactory classification of such a subject, but the following plan will be adopted:

1. The ordinary tumors in their order of frequency, as glioma, tuberculoma, cysts, and so forth, of the cerebellum itself.
2. Growths of the surrounding regions giving cerebellar symptoms, as of the fourth ventricle, medulla oblongata, pons, and corpora quadrigemina.
3. Growths in parts besides those mentioned, giving cerebellar symptoms.
4. Abscess of the cerebellum.
5. Internal hydrocephalus, with symptoms of cerebellar tumor.
6. Cerebellar symptoms without any lesions.
7. Lesions of the cerebellum without any symptoms.

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* From the Neuropathological Laboratory of the University of Pennsylvania.

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Excellent statistical studies of the frequency of the cerebellar and other cranial growths have been made, and without the desire to add to the already voluminous literature on the subject, it has been thought advisable to give a brief report of the brain tumors now in the neuropathological laboratory of the University of Pennsylvania, which is under the direction of Professor William G. Spiller. This collection has largely been accumulated in the last three or four years, and is from the services of Dr. Mills and Dr. Spiller, although in a number of instances specimens have been obtained from other sources.

Tumors of the cerebral cortex and subcortex	27
Cerebellar tumors	9
Tumors of the brain stem	9
Tumors implicating both brain and cord	4
Tumors of the cerebral cortex and subcortex:	
Sarcoma	13
Fibrosarcoma	2
Endothelioma	4
Glioma	3
Gumma	2
Carcinoma	1
Adenoma	1
Tuberculoma	1
Cerebellar tumors:	
Glioma	5
Sarcoma	1
Fibroma (in the cerebellopontile angle)	3
Tumors of the brain stem:	
Pons: Tuberculoma	2
Glioma	1
On the pons and medulla oblongata: Sarcoma	1
On the medulla oblongata: Chondrosarcoma	1
Within the fourth ventricle: Sarcoma	2
On the corpora quadrigemina: Fibroma	1
Within the corpora quadrigemina: Glioma	1
Tumors of the brain and cord: General sarcomatosis, with large tumors, especially in the cerebellopontile angle	4

Sections of at least seven other brain tumors were not considered, because definite knowledge of the location of the growths was lacking.

According to statistics tumors of the cerebellum are less frequent than of the cerebrum.

Schuster, in a statistical table of some thousand cases of brain tumor, found 21.6 per cent. to be cerebellar. When the relative size of the cerebellum and the cerebrum is considered, it is probable that new growths are more frequent in the former.

Tuberculous growths are more common in persons below the age of twenty years, while glioma, sarcoma, and cysts of various kinds are more frequent in the adult. The frequency of fibroma, especially of the acoustic nerve, is becoming better recognized. Syphilitic tumors of the cerebellum are rare. Of the other forms of new growths, as carcinoma, lipoma, angeioma, psammoma, and dermoid cysts, there are very few instances in the literature.

The lateral lobes of the cerebellum possibly because of their greater size, seem to be more frequently the seat of tumors than the middle lobe, although writers differ upon this point. Tumors within the middle cerebellar peduncle are rarely found, although a tumor within this peduncle is present in one of the specimens in the laboratory. Growths in the anterior and posterior cerebellar peduncles are also uncommon. The angle formed by the cerebellum, medulla oblongata, and pons is a favorite seat for new growths, these tumors growing either from within or upon the acoustic, facial, or trigeminal nerves, and frequently are fibromata.

Tuberculoma.—In 152 tuberculous brain tumors collected by Allen Starr, occurring in childhood, 47 were in the cerebellum. In the adult they are found with equal frequency in this region and in the pons and the cerebral cortex. They are nearly always multiple, and secondary to a tuberculous process elsewhere in the body. A tendency to symmetrical arrangement is also observed (Oppenheim). Their size varies from a small nodule to a large fist. Macroscopically, it is hard to distinguish a tuberculoma from a syphiloma. Both have poor blood supply and a tendency to caseate, the tuberculous growth to pus formation. Again, both have a tendency to grow from the meninges, although the tuberculous growths are found in the substance of the brain, and may have granulation areas and miliary tubercles about their border. It must be recalled, however, that syphilitic tumors of the cerebellum are rare.

The growth of a tubercle may be either rapid or slow. Tuberculous tumors may give no clinical symptoms. This has been explained by the slowness of the growth, the cerebellum gradually accommodating itself to increased pressure. Very recently, however, Raubitschek was able to demonstrate the persistence of the axis cylinders in tuberculous growths by Bielchowsky's method. This, as in multiple sclerosis, explains the persistence of function. Surgically, it is not advisable to operate upon these growths, as they are multiple and cannot be removed.

Glioma.—The cerebellum is a favorite seat for glioma. Five of our cerebellar growths were of such nature. Gliomata are almost always primary and single, although metastasis has been noted. The tumor may be as small as a cherry or as large as a hen's egg; it always grows from the brain substance itself, and is of slow growth. It is not sharply defined, but infiltrates into the brain substance, and it is difficult to tell it from normal brain tissue, although sometimes there is an increased consistence to pressure and there may be a slight swelling. The border zone of the tumor may present an increased number of blood vessels and there may be islets of new tissue.

Gliomata may be hard or soft, depending upon the excess of cells or fibrils, and have a yellowish white or reddish appearance. Cystic formation is very common, some authors believing that the whole tumor mass may disappear, leaving nothing but a cyst wall, and that it is necessary to examine microscopically the capsule to determine the gliomatous origin. Cysts form in the neighborhood of these tumors, and the surgeon may tap one of these cystic formations, believing it to be the only lesion present. It is wise, as Oppenheim has pointed out, to remove always a part of the cyst wall for microscopic examination. The fluid inside of these cysts may be whitish or bloody in character. Fatty, hæmorrhagic, and myxomatous changes occur in gliomatous tumors.

Microscopically, it is difficult to distinguish a glioma from a sarcoma unless a differential stain has been employed. There is some doubt as to the simultaneous occurrence of glioma and sarcoma, the so called gliosarcoma, some authors believing this to be impossible, as the former is of ectodermal and the latter of mesodermal origin. Others believe that by metaplastic processes a sarcomatous structure may develop from neurogliar tissue. According to certain pathologists, a gliosarcoma should only be diagnosticated where a sarcomatous, perivascular cellular mass is found within a glioma.

It can readily be understood from the slow growth and from its infiltrating character why clinical symptoms of brain tumor do not always appear, or not until late in the disease. Surgically, it is difficult or even impossible to remove completely such a tumor. Sections made from the specimens removed at the operation in Cases I and II of Dr. Mills and Dr. Frazier showed a glioma in each instance.

Sarcoma.—This form of brain tumor is about as common as the glioma, although in our experience sarcomata have been more frequently found. The growth may be small, flat, or nodular, or may be of large size. It is primary and usually solitary. Sarcoma always grows from the meninges, periosteum, or cranial bones, or from the pial covering of the blood vessels. It never grows from the brain substance, and therefore, unlike the glioma, it often compresses the brain tissue and may be distinct from it, although not infrequently it infiltrates the latter. Even when growing within the brain a distinct margin sometimes may be found, due to the softened area surrounding it. It is usually harder in consistency than a glioma, and is slow in its growth.

The tumor may soften or caseate. Myxomatous, hæmorrhagic, and cystic changes are not uncommon. Cystic changes are especially common in the cerebellum, not only in sarcomata, but also in gliomata. In one of Dr. Spiller's cases small sarcomatous masses were found in the walls of a cyst. If the fibrous tissue is very marked we have a fibrosarcoma.

Sarcoma may manifest itself as a diffuse multiple sarcomatosis. In an excellent article Spiller recorded two such cases and called attention to the rarity of this disease. He quotes Schlesinger, who subdivided the tumors under the head of multiple sarcomatosis into

(a) Diseases of the nervous substance and meninges,

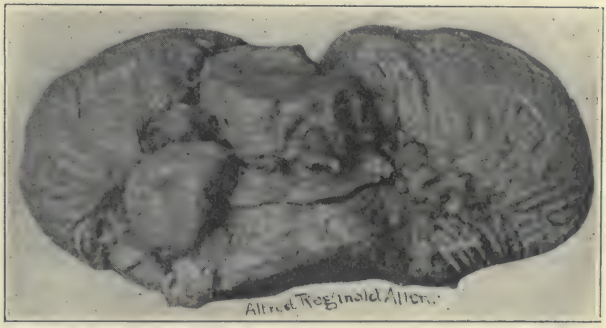

FIG. 1.—Sarcoma in left cerebellopontile angle. Small tumor in right cerebellopontile angle does not show in photograph.

(b) Multiple sarcomatosis of the membranes without sarcoma of the brain or cord, when it is (1) in the form of multiple small tumors, or (2) a diffuse sarcomatous infiltration of the membranes.

Of twenty cases recorded by Schlesinger, fourteen implicated the brain and cord or their membranes. " In nine of these fourteen cases cerebellar tumor was found, and in three the medulla oblongata was affected. It appears, therefore, that when the brain or its membranes are implicated in sarcomatosis, usually the structures of the posterior cranial fossa are affected, and that in about two thirds of the cases a tumor of the cerebellum is found."

In Spiller's first case a large sarcoma was found in the left cerebellar lobe, and in his second case a tumor was found in each cerebellopontile angle, the larger one being on the left side, as shown in Fig. 1. Tumors were also found in this case in the Gasserian ganglia, pituitary body, floor of the fourth ventricle, right internal auditory meatus, and right jugular foramen, and numerous small tumors were found in the pia of the spinal cord.

It may be impossible, as in Spiller's second case, to make a correct diagnosis in sarcomatosis of the brain and of the pial covering. Extensive alteration may cause few clinical symptoms, because the soft tumor masses grow in the pia and about the cranial nerves and spinal roots, and may produce little or no compression or destruction of the nervous tissue. Spiller insists upon the importance of remembering this fact, for when evidences of sarcomatosis are found, the case is an inoperative one.

Occasionally the process may invade the brain substance, while the nerve roots may escape. It is, according to Spiller, because of this escape of the nervous tissue in many cases that a correct diagnosis of the extent of the process may be impossible.

The infiltration of the pia may resemble that caused by syphilis or tuberculosis. Again, as in Nonne's case, the macroscopical examination may be normal.

Sarcomatous tumors, according to Westphal, occur more often in the young. When tumors occur in the posterior cranial fossa they have a predilection for the cerebellopontile angle and the internal auditory meatus.

Isolated sarcomata whether of the cerebellum or of any other region of the brain are, next to fibromata, among the most favorable forms of tumor for surgical removal. Of course the question of multiple sarcomatosis must always be carefully considered when deciding upon operation. With regard to surgical procedure the hard non-infiltrating sarcomata are the most favorable. Experience shows, however, that a sarcoma which appears to be infiltrating when the brain and tumor mass are first exposed, is often separable from the brain substance.

Syphilitic Growths.—Gummata are rarely found post mortem, although they are possibly the most common cranial growths. They are especially rare in the cerebellum. The resemblance between this growth and tuberculoma has already been discussed. In a recent article Mills recorded two cases in which the diagnosis of a tumor in the cerebellopontile angle was made. At the necropsy no tumors were apparent, but microscopically in the first case a diffuse syphilitic basal meningitis was found and in his second case besides a meningitis at the base, there were numerous areas of softening throughout the brain, extending from the gray into the white matter. These areas of softening were yellowish red in color, soft in consistence, and were well defined from the surrounding brain substance. Microscopically, there was an intense round cell infiltration about the blood vessels and within the tissues.

These cases illustrate well the nature of syphilitic new growths. It is well known that a syphilitic basal meningitis, or meningoencephalitis, may attack any cranial nerve or combination of cranial nerves, but according to Mills they show a predilection in favor of the nerves from the second to the seventh inclusive, of these the fifth perhaps most frequently escaping.

Syphilitic growths are rapid in development, but it must be remembered that the various pathological conditions which lead on to these growths have been long present.

Fibromata.—These tumors are rare, but they are relatively more frequent in the cerebellum than in the cerebrum, and especially in the cerebellopontile angle. This has been better recognized within the last few years, because of the relatively successful surgical removal of tumors growing in this area.

A fibroma invading the cerebellopontile angle may be only a part of a general neurofibromatosis; this, however, is rare, or, what is more common, it may be the only expression of this process, a central neurofibromatosis. The growth is slow, and generally is unilateral, although in rare instances it may be present on both sides. Henneberg and Koch pointed out that these tumors are more often found on the left side in the ratio of three to two. In the cases reported by Dr. Mills, the pathological reports of which are here given, the neoplasms were on the left side. In an examination of the tumors situated in the pons, medulla oblongata and the cerebellum, we found that the majority were on the left side. It seems, therefore, that tumors of these areas are more prone to grow on the left side.

The fibroma may be as small as a cherry or the size of a large egg. The growth is firm, hard, nodular, and has a distinct capsule surrounding it. It is loosely attached to the brain by an atrophic nerve trunk, a few blood vessels or a meningeal process, and these attachments may be easily ruptured. These tumors are in organic relation, especially with the acoustic nerve, and more rarely with the trigeminus and facial nerves. They nearly always grow from the endoneurium and rarely from the peri- or epineurium. Consequently we may find medullated nerve fibres either in the periphery of the tumor or in its centre. As a rule, if the process involves the other cranial nerves, we have a general neurofibromatosis.

The fibroma may undergo a cystic, fatty, or myxomatous degeneration. Very often in its advanced stages it may assume a sarcomatous tendency. Histologically we find a connective tissue structure with entire absence of nerve elements, except sometimes a few medullated nerve fibres either in the periphery or its central part. These are remnants of the nerve on which the fibroma grows and should not be mistaken for a part of the new growth. Most writers persist in calling these tumors neurofibromata. The best example of a true neurofibroma is the amputation neuroma, therefore, a fibroma would be a better term for these growths.

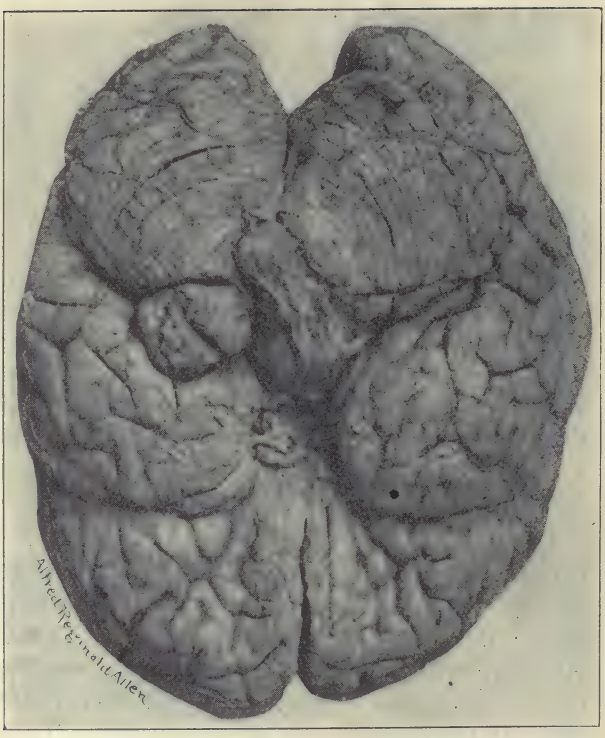

FIG. 2.—Fibroma growing from the left acoustic nerve compressing slightly the left lateral lobe of the cerebellum and the lower surface of the left temporal lobe.

In a number of cases of fibroma of the acousticus there were associated cortical changes. Henneberg and Koch reported hyperplasia and hypertrophy of the glia cells of the cortex, especially of the deeper layer, and in another case endothelioma and psammomata of the dura mater. Fraenkel and Hunt made a similar observation. In another case reported by these authors there were protrusions and minute herniæ attached to and sometimes perforating the dura. Histologically, these consisted of large cells of the spindle type and of glia cells.

At times the fibromatous process may involve the whole of the intracranial portion of the acousticus. In a case of Alexander and v. Frankl-Hochwart, an anatomical examination of the labyrinth showed a degenerative atrophy of the cochlear nerve, the spiral ganglion, the organ of Corti, and the striæ vasculares.

These tumors compress greatly the lateral lobes of the cerebellum, the pons, and the medulla oblongata. In one of Dr. Mills's cases the temporal lobe was compressed. Because of the slow growth and the nature of the tumor, clinical symptoms may not appear at all, or only late in the disease. In one of Dr. Mills's cases there were no symptoms of such a growth, the tumor being found at necropsy.

The following pathological report of two cases of fibroma in the left cerebellopontile angle are from the service of Dr. Mills. They are referred to by him in the discussion of tumors of the cerebellopontile angle, the first extensively, the second briefly. The clinical report of the second case is appended:

Case I.—A tumor (Fig. 2) 3 cm. wide and 2½ cm. in length anteroposteriorly was found in the left cerebellopontile angle, compressing slightly the forward part of the left lateral lobe of the cerebellum and the under surface of the temporal lobe. The pons and medulla oblongata were not compressed. There was marked internal hydrocephalus. Microscopical examination showed it to be a fibroma.

Case II.—A tumor (Fig. 3) 5 cm. wide and 3 to 4 cm. in length anteroposteriorly was found in the left cerebellopontile angle. The tumor was very firm and nodular, and compressed considerably the under surface of the left lateral lobe of the cerebellum and the left side of the pons. At no place did the tumor infiltrate the brain tissue, and it could be entirely enucleated. Microscopical examination showed it to be a fibroma.

Cysts.—Cystic degeneration of gliomata and sarcomata are very common, especially if they are situated in the cerebellum. This has been discussed when speaking of sarcoma. Other tumors, as fibroma and carcinoma, are prone to undergo cystic change, but more rarely. Some authors believe that the whole tumor may disappear and only a cyst remain. In other cases only a microscopical examination will detect a small tumor mass in the walls of the cyst. Spiller has pointed out that the wall of a congenital cyst may be the starting point for a neoplasm, and this possibility should not be ignored.

The most common cystic changes found in the brain are due to parasitic growth, the cysticercus cellulosæ and the ecchinococcus. These, however, are so rare in this country that they will not be here discussed.

Cysts due to traumatism are recorded, but their genesis is by no means clear. Congenital cysts are rare. They are probably offshoots of the primary cerebral vesicles. Dermoid cysts

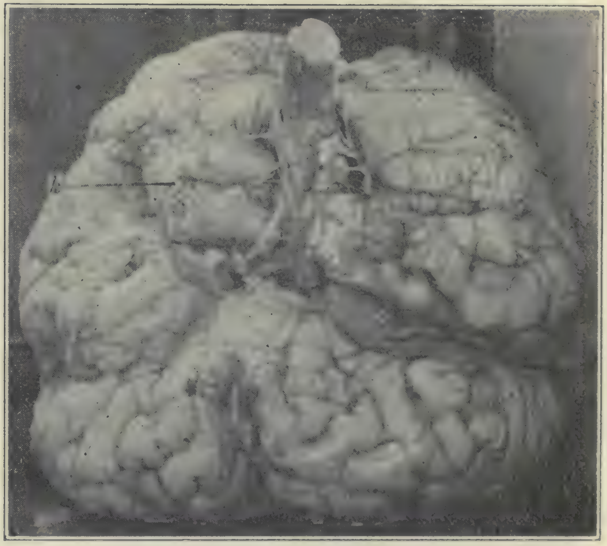

FIG. 3.—Fibroma growing in the left cerebellopontile angle compressing the lower surface of the cerebellum and the left side of the pons.

have been recorded as occurring in the cerebellum in several instances.

Carcinoma.—Carcinoma of the cerebellum is rare. This form of neoplasm is always secondary and grows from the dura or in the substance of the brain. Saenger recorded infiltration of the cerebral pia with cancer cells. The possibility of toxic changes must be considered, as it is not improbable that through intoxication caused by a carcinoma elsewhere in the body, symptoms of brain tumor may be present.

Osteoma.—In several instances an osteoma has been described as occurring in the cerebellum. It is probable that these growths are not primarily of bone formation, but are the result of calcification of such tumors as tuberculoma, fibroma, sarcoma, and even lipoma. Other neoplasms, as adenoma, lipoma, angeioma, psammoma, and cholesteatoma are hardly ever found in the cerebellum, so they will not be discussed. It must also be remembered that aneurysm of the vertebral or basilar artery may give symptoms of cerebellar growth.

The Influence of Cerebellar Growths.—At the operation when the dura is removed there is nearly always increased tension and the parts may bulge. The surface of the cerebellum is flat and the fissures may be abolished. The pia covering the neoplasm is generally poor in its blood supply. The tissues near the growth may be softened. If the tumor is in the lateral lobe of the cerebellum it may compress the fifth, seventh, and eighth cranial nerves. The occipital lobes may even be compressed through the tentorium. If the cerebellar tumor is large it may compress the corpora quadrigemina, pons, and the medulla oblongata, and these structures may be flattened or deformed. Pressure may also be exerted upon the cranial nerves at the base of the brain. The influence of cerebellopontile growths upon surrounding structures has already been discussed.

The cerebrospinal fluid is almost always increased in cases of cerebellar tumor, because pressure is exerted upon the communication between the lateral ventricles and the fourth ventricle, or upon the veins of Galen, which convey the blood from the chorioid plexus to the sinus rectus. Because of this internal hydrocephalus undue pressure is brought to bear upon the different cranial nerves, as the optic and olfactory. The optic chiasm may be directly compressed through pressure from the third ventricle.

Alterations in the posterior roots and the posterior columns of the spinal cord have been recorded as occurring in conjunction with tumors of the brain. Such changes have been also found by Dr. Spiller. According to Batten and Collier they are especially present in cerebellar growths, and are due to the increased pressure. Dinkler and Becker believe that toxic or nutritional changes are at fault.

Tumors of the Fourth Ventricle, Medulla Oblongata, Pons, and Chorioid Plexus.—It is not in the province of this paper to consider *in extenso* neoplasms of these areas, but inasmuch as these growths sometimes give symptoms of cerebellar involvement, they will be briefly considered.

Tumors of the fourth ventricle and of the medulla oblongata may give no appreciable clinical symptoms. They may either be cystic or hard, and may grow in the substance of the medulla oblongata. This is especially true of parasitic and congenital cysts. Hunt recorded two congenital cysts of the fourth ventricle in which the cerebellum was greatly compressed and yet there were no cerebellar symptoms.

Neoplasms growing within or upon the corpora quadrigemina nearly always compress the middle or the lateral lobes of the cerebellum. They also cause internal hydrocephalus. Two such specimens are in our collection.

Tumors of the pons may cause pressure symptoms upon the cerebellum, or the growths may involve the middle cerebellar peduncles. Growths of the chorioid plexus, as in a case of Arnold's, where a psammoma of the size of an apple was found, may compress the pons, medulla oblongata, and the cerebellum.

Tumors in the Cerebrum Giving Symptoms of Cerebellar Growth.—Ascherson recorded an instance in which a sarcoma was found in the centrum ovale of the left side in the upper motor area. This neoplasm measured 1½ by 2⅜ inches, and could easily be enucleated. It caused a compression of the lateral ventricle in the same side. This author cites Raymond as having recorded an almost similar case. Ascherson is of the opinion that the cerebellar symptoms were due to pressure exerted through the lateral ventricle, and he emphasizes the importance of early symptoms before those of pressure are apparent. In this connection the fact that tumors of the postparietal cortex or subcortex, may give unilateral ataxia should be borne in mind. The diagnosis between post parietal and cerebellar tumors is given in the paper of Dr. Mills.

Abscess.—Chronic otitis media is the most frequent cause of abscess in the cerebellum. It may be due to such other causes as traumatism or may be a part of a general pyæmic process, but these instances are uncommon. The abscess occurs mostly in the anterior outer part of the cerebellum, and is generally single. It may involve also the adjoining temporal lobe. The abscess may be encapsulated or it may keep on forming pus. Surrounding it, œdema and softening of the brain substance are found. Pus may travel along the facialis and acousticus, and cause extradural abscesses. Hydrocephalus, sinus thrombosis, and thrombophlebitis are frequent complications.

Internal Hydrocephalus.—This condition is most often caused by a brain tumor, but it may be congenital or acquired. Spiller recorded an instance in which the symptoms were those of cerebellar tumor, and at the necropsy the cerebral ventricles were much distended, but the fourth ventricle was of normal size. The aqueduct of Sylvius was almost entirely occluded when examined, and the occlusion must have been congenital or have occurred early. Byrom Bramwell recorded a similar case, but here a localized meningitis caused a closure of the foramen of Magendie. The possibilty of internal hydrocephalus should always be kept in mind when a cerebellar growth is considered.

Symptoms of Cerebellar Tumor Without Lesions.—In a very important paper, Nonne called attention to those cases in which the majority of the symptoms of brain tumor were present, and in which either spontaneously or under mercurial treatment the symptoms disappeared, leaving, perhaps, a partial optic nerve atrophy. There was no reason in any of the eight clinical cases he reported to suspect syphilis.

He also records three similar cases with necropsy, in two of which symptoms of a tumor in the posterior cranial fossa was diagnosticated. At the necropsy in the first case, internal hydrocephalus was found. On the floor of the fourth ventricle, opposite the posterior medullary velum, there was a hard, long, yellowish white structure, which obstructed the flow of the cerebrospinal fluid. Microscopically this was found to be a fibroma. In his second case, internal hydrocephalus was also found, this being caused by a sarcoma of the ependyma of the floor of the fourth ventricle.

He further records three cases with necropsy in which there was no internal hydrocephalus. Nonne also reports cases of internal hydrocephalus which gave largely basal symptoms due to various causes, and which terminated either in death or recovery. He leaves us in doubt as to what is the cause of such a condition.

Dr. Spiller has very kindly given me the records of such a case occurring in his service.

Woman, 44 years of age, domestic, past history unimportant. Two and one half years ago the patient began to have violent headache in the left cerebellar region. This headache became more severe and became localized in the left parietal region, where there was also great tenderness to pressure. She had an ataxic, drunken gait, and would fall to the left or backwards when walking. Extreme vertigo was also present, especially when she was lying on her left side or walking. Power was diminished in the lower limbs and sensation was also somewhat impaired. The patellar jerks were absent. There was no albumin in the urine.

Dr. Roberts operated at the point of great tenderness in the left posterior parietal region. There was nothing abnormal found and the brain appeared to be in a healthy condition.

The patient's symptoms steadily disappeared, the pain in the head became better and in a short time she seemed almost well.

Another case was studied by me repeatedly.

This woman was in the nervous wards of the Philadelphia General Hospital in the service of Dr. Spiller. She was 52 years of age, denied venereal history, and her past history was unimportant. Five years ago she began to have violent vertical headache, which has persisted more or less since. One year ago she began to have objective vertigo, and convulsions, Jacksonian in type, which always involved the left side of the face, and the left arm and leg. Sight also became poor at that time, and her memory was not as good as formerly. In my examination she showed a paresis of the left arm and leg, these being spastic, and the reflexes were exaggerated. The Babinski sign was present on this side. There was also a paresis of the lower distribution of the left seventh nerve, and a paralysis of the left abducens and the left fifth nerve, both in its motor and sensory distribution. Optic neuritis was present in both eyes.

On protruding her tongue she had a clonic to and fro movement which became apparent on talking or moving the tongue. She became steadily worse and finally was comatose. The urine examination was negative. She rallied, however, her symptoms steadily disappeared and she was discharged from the hospital four months afterwards, the only remaining symptom being a dimness of vision.

These two cases are similar to those recorded by Nonne. No adequate explanation for them can be given.

Lesions of the Cerebellum Without Symptoms.—These lesions may be either congenital, acquired early in life, or may be tumors. It is not surprising that injuries to the cerebellum early in life or that tumors of slow development which occur in the same period give no appreciable symptoms, because the functions of the cerebellum in such cases have probably been assumed by other parts of the brain. Lesions of the lateral lobes of the cerebellum are less liable to cause symptoms than when they implicate the entire cerebellum. Spiller recorded three cases of lesion of the cerebellum in which there were no symptoms, and he also reviewed the literature upon this subject. In his first case one cerebellar lobe was smaller than the other, and it was sclerotic. In the second case there was a tumor upon the corpora quadrigemina in which the lateral lobe of the cerebellum was compressed. The third was one of tumor within the vermis.

Cases are recorded in which tubercles involved an entire lateral lobe and gliomata and cysts occupied the middle lobe, and yet there were no symptoms. Oppenheim refers to a case of Putnam's, where the only symptom for years was an optic nerve atrophy, in which at necropsy a cyst of the cerebellum was found. He also refers to Bramwell's case, where in a thoroughly studied case no symptoms were apparent, while at necropsy four tumors were found.

It can readily be understood why symptoms may not be apparent in a gliomatous tumor, because of its infiltrating character, and in tubercles, in which the axis cylinders are retained; but it is difficult to explain the absence of symptoms in the other instances.

The notes of the case of cerebellopontile tumor, as shown in the illustration, Fig. 3, and Case II of this paper, were furnished by Dr. Mills. The patient was seen by Dr. Mills in consultation with Dr. W. W. Keen; she was also examined in consultation by Dr. W. G. Spiller. The tumor sprang from the eighth nerve, and the chief focal symptoms were one sided deafness, tinnitus, facial monospasm, hypæsthesia of one side of the face, nystagmoid movements, slight paresis of right abducens, and vasomotor and cardiac disturbances. Severe headache, nausea, vomiting, and optic neuritis were also present.

This patient was a married woman, 30 years of age, five of whose maternal relatives had died of cancer. Four years before coming under observation the ossicles of her left ear were removed, on account of an annoying tinnitus, but without the desired result. About one year later she began to suffer from severe headache. The next year slight optic neuritis was observed in both eyes, the neuritis going on to atrophy and blindness, which was complete in less than two years. Headache, nausea, vomiting, and depression were recurring symptoms, and taste and smell were impaired. During two or three years she was treated for various complaints as anæmia and neurasthenia, and both Graves's disease and interstitial nephritis were suspected. About six months before coming under observation she had a convulsion with loss of consciousness, this being followed by several others of a similar kind.

The patient was having at somewhat frequent intervals attacks beginning with pain in the head, which was referred to the forehead and eyes. In these she became nauseated and then vomited, becoming pale or even cyanosed, with loss of consciousness. The vomiting was preceded or accompanied by marked facial monospasm, in which the mouth was drawn forcibly to the left and the eyelids were drawn together. Only the left side of the face was involved in the seizure. Examination showed that she had no ataxia of station or gait. Hearing on the right side was good, on the left side it was abolished. The mouth deviated slightly to the left when opened widely. Hypæsthesia to pain was present on the left side of the face and head, and sometimes appeared to be present in the left hand. The patient was not mentally impaired, but was easily exhausted mentally, and was at times irritable and depressed as the result of her sufferings. During the time she was under observation she had frequently recurring headaches, usually severe, sometimes accompanied with nausea or even vomiting, and sometimes with the facial spasm already described. Irregular nystagmoid movements occurred when the patient turned the eyeballs to the extreme right or left. She had complete loss of smell, and loss of taste on the left side of the tongue. On one occasion it was thought that the facial spasm was accompanied by some spasmodic movements of the left hand, but this was doubtful, and even the observer thought it may have been a voluntary movement.

The question of the existence of exophthalmic goître was one which arose for diagnostic discussion several times during the history of this case. The diagnosis of this affection was first made a year or two before coming under our observation. It was also considered and favorably regarded by some of those who saw her in consultation late in the case. Her eyes had somewhat the staring expression of the blind; they were rather large, but her relatives stated that she had always had prominent eyes, and the exophthalmos was apparent rather than real. The enlargement of the thyreoid was so doubtful as to cause some disagreement among those who examined her as to its existence. A slight enlargement of the gland on one side seemed sometimes to be present. Her pulse frequently, perhaps usually, was between 100 and 110, and sometimes rose above the latter point. It was a pulse such as is not infrequently seen in the late stages of an exhaustive intracranial disease. Graves's disease was finally excluded. In the light of the post mortem findings, it is not improbable that some of the symptoms simulating this affection were due to the tumor, from its position, causing vasomotor and cardiac disturbances.

Ocular and ophthalmoscopic examinations were made by Dr. W. C. Posey, who reported as follows: Ocular movements good in all directions, except externally to the right, where there is a slight limitation of movement, the right eye not being brought as far as normal into the external canthus. On fixation in the median line and below, the eyeballs are quiet. Marked lateral nystagmic movements appear, however, as soon as the eyes leave these primary positions, the nystagmus being most marked on extreme outward rotation to the right and to the left. The pupil in the right eye is round, and is 5 mm. in size; that in the left eye is oval, 3 by 4 mm., with its long axis at 50°. The irides do not respond to light or accommodation stimuli. The ophthalmoscopic examination reveals clear media in each eye, with the signs of regressive optic neuritis. The swelling of the nerves, however, is still very marked, both papillæ projecting into the fundi to the extent of 2 or 3 mm. The nerves are gray and succulent looking, and the retinal arteries and veins are tortuous and cord like. One nerve is not more swollen than the other. There are no extravasations or hæmorrhages, or traces of either of these in the fundi. The patient is totally blind.

Other examinations were made by Dr. Posey, but they did not demonstrate anything different from what is above recorded.

Eventually an operation was performed in this case by Dr. W. W. Keen, by whom the patient was seen in consultation with Dr. Mills. Although a tumor at the base was considered, it was thought for several reasons that the lesion was probably in or beneath the facial centre. In the first place sufficient consideration was not given to the tinnitus and deafness. Owing to the fact that a peripheral operation had been performed early for the relief of the latter, it was supposed that the impairment and disturbance of hearing were due to causes which were at least in part peripheral. The facial monospasm was much like that which is observed in the case of subcortical or cortical growth. It is interesting to note that some disease of the cortex was present at the position of the trephining, as demonstrated at the necropsy, but no tumor was found here. The patient died a few hours after the operation.

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THE TREATMENT OF CEREBRAL PALSIES AND ATHETOSIS BY NERVE ANASTOMOSIS AND TRANSPLANTATION.[1]

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REMARKS BY DR. SPILLER.

The views presented in this paper are put forward with much hesitation, because the means by which voluntary power is restored in hemiplegia are unknown, and the conditions have resisted most attempts at improvement. The views here expressed are original with the authors, and if experience should prove that they are fallacious, they may nevertheless afford some suggestions for further investigations. We should have preferred to wait until these investigations had been carried to a successful conclusion before making any reference to them, but it may be better to invite discussion on the subject.

Is it possible to benefit in any way by surgical means the patient who is afflicted with an incomplete hemiplegia? This is a question I have repeatedly proposed to myself. Where the hemiplegia is complete or nearly complete we can not hope to accomplish anything by any surgical procedure. There are, however, many cases in which there is a partial return of power in the paralyzed limbs, and experience shows that usually the restoration of motion is greater in the flexors in the upper limb and greater in the extensors in the lower limb, except those of the toes. I have—to give one example—under my observation at present a man, aged twenty-seven years, who has had left hemiparesis so long as he can remember. He has excellent power in the flexors of the hand, but can extend the hand and fingers only to the plane of the forearm, and he has no contractures. In the lower limb he can not extend the toes or dorsally flex the foot, although he can extend the foot with much power, and in walking raises the left knee a little more than the right, because the left lower limb is too long. An operation on the upper limb would not be permissible, because the flexors of the fingers are more useful than the extensors, and he can extend the fingers to a level with the back of the hand. In other cases, however, the difference in the degree of power of the flexors and extensors of the fingers and hand is greater, and the restoration of power may occur almost alone in the flexors. The latter may be contractured so that the hand is flexed on the forearm and the fingers flexed into the palm. The restoration of the power of the flexors of the fingers and hand may be of little or no value, unless there is at least a partial return of power in the antagonistic muscles, although this return need not be so great in the extensors as in the flexors. I have thought that in a case where the flexors alone regain power we might anastomose the central ends of some of the least important of the flexor nerves with the peripheral ends of extensor nerves, and in this way restore more nearly the normal relation between the flexor and extensor muscles. In the case I have referred to above, in which the flexors of the toes were much stronger than the extensors, an anastomosis of the central ends of one or more flexor branches with the distal ends of extensor nerves might be of decided advantage. Even though the hemiparesis has existed many years, regeneration of the united nerve ends would occur, because these nerves still have power and their connection with their spinal cells of origin has never been impaired. The muscles of course must not be entirely wasted.

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[1] Read by invitation before the New York Neurological Society, March 7, 1905.

From the Laboratories of Neuropathology and Surgical Pathology in the University of Pennsylvania.

The question now arises, would impulses pass from the brain over the central motor tracts to the anastomosed fibers in such a way that useful return of function might be expected? I think the probability is that such a restoration in part at least would occur. It is true that movements and not muscles are represented in the brain cortex, but it has been demonstrated that when the lesion is in the peripheral nerves and anastomosis of nerves has been performed, a new form of associated movements may be learned by the brain.

We need not enter into a discussion of the manner in which return of function occurs after hemiplegia. Whether there is a close association in the brain of the fibers for the agonistic and antagonistic muscles, to use the terms employed by certain German writers, as Mann believes, need not concern us now. The partial return of power after a hemiplegia, according to the view of Rothmann, may be the result of independent function newly acquired by subcortical centers, especially by the optic thalamus, and is such as exists normally in lower animals, or is developed in the monkey a few days after destruction of the cortical centers for the limbs.

It is certain that for perfect voluntary motion we must have return of power in both agonistic and antagonistic muscles. We can not flex a finger properly unless the extensor of that finger is in the requisite degree of tonicity. If we flex the fingers of one hand firmly into the palm and place the other hand over the extensors of the wrist, we find that the extensors of the carpus become tense. It is in large measure because of this loss of extensor power, and thereby of fixation of the hand, that flexion of the fingers is weakened when we have wrist drop from a lesion of the posterior interosseous nerve.

In order to study the conditions of paralysis existing in hemiplegia I have examined twenty-six cases in which the paralysis dated from early childhood. Dr. Martin W. Barr kindly had collected for me all the cases of infantile hemiplegia at present in the Pennsylvania Training School for Feeble-Minded Children. I wish to thank Dr. Jennings and Dr. Weisenburg for their assistance in the study of these cases.

The return of power is, I think, greater on the average in cases in which the hemiplegia develops in childhood than in those in which it develops in adult life. These twenty-six cases were not selected from a large number of hemiplegics, and yet in eleven of these the voluntary power in the flexors of the hand and fingers was much greater than in the extensors; and in four of these the voluntary power of the flexors of the toes and extensors of the foot was much greater than in the extensors of the toes and flexors of the foot. The cases in which the hand had regained partial power were more numerous than those in which the foot had regained partial power. In eleven cases out of twenty-six, therefore, the flexors of the hand were so much stronger than the extensors that a nerve anastomosis might properly be attempted, but only four cases offered much hope for improvement from an operation on the foot. All cases in which the restoration of power in the extensors of the hand and fingers was fair, even though less than in the flexors, were rejected as being undesirable for operation. As one of the most suitable cases the following may be briefly described:

L. A., female, aged thirty-one years, high grade imbecile, has voluntary movements at all parts of the right upper limb, but the movements are impaired. The fingers of the right hand are slightly flexed into the palm. She has considerable power in the flexors of the right hand, but no power in the extensors. In making a bed she is able to hold the bed clothing with her right hand, but the use of the hand is much diminished by the permanent flexion. The paralysis in the muscles of the right foot is almost complete.

There is probably therefore a field for surgical intervention in certain cases of cerebral hemiplegia, but the cases must be carefuly selected, and each thoroughly studied. Whether any benefit by nerve anastomosis will result to the hemiplegic individual or not, experience alone will decide. The subject, however, is one deserving attention. Dr. Frazier has in one case operated upon the hand of a partially hemiplegic woman, but the return of power following the hemiplegia has not been sufficient until the present time to make the intervention entirely satisfactory. He will describe his method of anastomosing the nerves. He has also made investigation upon dogs concerning the results of anastomosing flexor nerves with extensor nerves.

Tendon transplantation for the increase of function in paralysis of all varieties has proved of benefit provided the paralysis is not progressive,—a subject upon which Oppenheim has written within the last few weeks—but where nerve anastomosis or transplantation is successful we have a means of restoring the function of muscles previously paralyzed, and therefore this method is better than tendon transplantation which makes no demand on paralyzed muscles.

I am well aware that improvement may occur in hemiplegia years after the paralysis has first developed.

Athetosis is one of the most distressing forms of involuntary movement, and the failure to control it in any degree is a reproach to medicine. I have for several years been trying to devise some method of treatment for this condition. Much study has been given to the causation of these movements, but our knowledge regarding the condition is very insignificant. There must in cases in which athetosis exists be an irritation of the motor system somewhere, and the investigations of recent years (Bonhoeffer, Mann) seem to point to implication of the superior cerebellar peduncle. We can not hope to remove the irritation in the brain. We can not hope to cut the central motor fibers. Such a procedure would be unjustifiable. Can we accomplish anything by operation upon the peripheral nerves?

Where athetosis exists the muscles are in a state of greatly increased tonicity and often are abnormally well developed and abnormally strong. Theoretically the proper procedure might be to cut posterior roots of the affected limbs, the number to be cut depending on the condition in each case; but this is always a serious operation, and the results have at times been unexpectedly grave.

It is possible that if we were to divide one or more of the motor nerves of the affected limb and immediately suture the divided portions we might lessen the involuntary activity and weaken the muscles only slightly; or if experience taught us to make the paresis greater by making the conditions for full restoration of power less favorable, we might be justified in leaving some of the nerves divided, or even in making reunion of certain of the nerves improbable. It is possible that in this way we might diminish the nervous impulses sent out continually and involuntarily from the brain, or at least check their manifestation. In athetosis the flexors of the hand and fingers are often unusually strong, and where this is the case the union of the peripheral end of a flexor nerve with the central end of an extensor nerve and *vice versa*, might restore the proper relations of voluntary power and lessen or abolish the involuntary movements. We have at present a case of marked flexor spasm with athetosis on which Dr. Frazier will operate.[2]

Nerve transplantation if the lesion is in the spinal cord is a hopeful field for surgical intervention when the conditions are favorable. I[3] have already discussed this subject in association with Dr. Frazier, in a paper read before the Section on Nervous and Mental Diseases of the American Medical Association in June, 1904, and may say here that paralysis confined to one or a few muscles is not an uncommon occurrence in anterior poliomyelitis, and that the cases in which nerve anastomosis may be done are therefore fairly numerous.

REMARKS BY DR. FRAZIER.

The adaptation of the operations known as nerve anastomoses or implantations, to the treatment of cerebral palsies, as proposed by Spiller, opens up a field in neurological surgery which seems to me to be full of promise. There are innumerable details in the method of executing the operation, which can be determined only by thorough and repeated experimental investigations and clinical observations. A series of experiments is now being conducted by us, but few have as yet reached the stage which warrants their publication. Our first clinical experience dealt with a woman 65 years of age, who five years ago had an apoplectic attack. When the operation was performed, July 28, 1904, she was hemiplegic; there was marked atrophy of the muscles of the hand, forearm and arm, but the paralysis was so much more marked in the extensor than in the flexor group of muscles that she could partially, and this only with great difficulty, extend the proximal phalanges. Her hand was a greater part of the time held in partial flexion. Under ether anesthesia an incision was made along the inner border of the coraco-brachialis, exposing the brachial vessels and the main nerve trunks. The median and musculo-spiral nerves were isolated sufficiently for our purpose and a flap, composed of one-half of the median nerve, with its base centralwards, was implanted into the musculo-spiral. Union was effected by making a longitudinal incision in the sheath of the musculo-spiral into which the free end of the flap from the median nerve was introduced and sutured in such a way that the nerve fibers of the median were imbeded into those of the musculo-spiral. The results of the operation up to the present time have not been very positive although it is still possible that as time goes on greater improvement will be noted. There is probably distinct improvement in the extension of one of the fingers. It is too early yet to predict the result.

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[2] Since this paper was read nerve transplantation has been done and the patient's condition is distinctly improved.

[3] Journal of the American Medical Association, Jan. 21, 1905.

The experimental work is being carried out on dogs and has consisted essentially in anastomosing the nerves presiding over one group of muscles with those of another. Thus for example, in several instances the peripheral end of the divided sciatic nerve has been joined to the central end of the divided anterior crural, and the peripheral end of the anterior crural with the central end of the sciatic. In these cases the operation was followed by transitory paralysis, but during the third week the animals began to regain power in the affected limb. At first in their attempt to walk the affected limb was extended with a jerky movement, instead of being flexed as it should, and after three or four attempts at using the affected limb the animals apparently became discouraged and ran around on three legs. About a week later the animals had apparently learned to control the limb, at least when walking, although in attempting to run they again became confused and the limb would be extended, when it should have been flexed. Before two months had elapsed, however, these incoordinate movements had disappeared.

These experiments are cited simply as illustrating the lines along which the experiments are being conducted. Many modifications of this plan will of course have to be worked out in detail before the experiments will be reported in full. As compared with tendon transplantation, the only other surgical procedure which can be considered as an alternative, nerve anastomosis, theoretically at least, is the more rational measure. If one is able to restore function to the paralyzed muscle, the functional results should be better than if an attempt is made to compensate for the loss of function of one group by putting an additional tax upon another.

Applied to the treatment of those palsied conditions with uneven distribution of power, in those muscles, which oppose one another, I cannot but feel that in the transference of a proportion of nerve force, in an amount still to be determined, from one group of muscles to another, from the stronger to the weaker, we have at our disposal a means of control which may be productive of gratifying results.

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PERIPHERAL OBLITERATING ARTERITIS AS A CAUSE OF TRIPLEGIA FOLLOWING HEMIPLEGIA, AND OF PARAPLEGIA.

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We purpose to describe a not very infrequent but much neglected form of palsy caused by obliterating arteritis in the extremities affected. When it affects the arteries of the legs in a hemiplegic the result is a triplegia which may be thought to have been caused by cerebrospinal disease if the possibility of local vascular disease and the disability resulting therefrom are not thought of.

During the acute stage of a sudden hemiplegia caused by cerebral hemorrhage or thrombosis, but never in the slowly oncoming palsy from cerebral tumor, there is, on account of the bilateral control of movements in the cerebral cortex, a temporary lessening of power of the arm and leg on the same side as the lesion. It is also well known that in the chronic stage of hemiplegia the deep reflexes on the non-paralyzed side are often permanently increased and that even true ankle clonus may be present. This temporary partial palsy and permanent exaggeration of the deep reflexes arise without any disease of the brain and cord except that which caused the primary palsy. Occasionally, however, the disturbance of function of the leg, which on theoretic grounds should not be affected, is far greater than this, and it becomes as powerless as the other, both thighs being flexed on the abdomen and the calves on the thighs, the final result being a triplegia producing a clinical picture superficially resembling that seen when diffuse myelitis has occurred in a hemiplegic. The condition must not be mistaken for the triplegia which sometimes occurs in syphilis as the result of multiple cerebral and spinal lesions, nor for double hemiplegia or hemiplegia plus monoplegia resulting from bilateral cerebral lesions. It slowly follows a single cerebral apoplexy and is caused, as is shown by post-mortem examination, not by involvement of the spinal cord, but by disease of the arteries of the legs themselves.

The condition does not seem to be very common when the frequency of hemiplegia is considered. It never follows embolic hemiplegia in young people, but occurs only in the aged whose arteries are diseased throughout the entire body.

The history of the case we report is as follows:

W. S., a white man, aged seventy-eight years, was admitted to the Philadelphia Hospital November 16, 1904, with right-sided hemiplegia. His past history is of no interest except that he had had four apoplectic strokes. The last, and the one for which he desired admission to the hospital, occurred about six months before. While standing he suddenly fell unconscious and when consciousness returned was found to be palsied on the right side of the body and speechless. He did not improve in speech or motor power, was weak mentally, and lost control of the bladder and rectum.

Examination. A greatly emaciated old man with spastic paralysis of the right arm and leg. He was completely aphasic, neither speaking nor understanding speech. He made no effort to express thought by pantomime. The reflexes were noted to be increased in each leg, but nothing was said in the notes of this examination as to any loss of power in the left leg. The right pupil was a little larger than the left and both reacted sluggishly to light and convergence. All the peripheral palpable arteries were much thickened. He was bedridden.

When we saw him in January, 1905, both thighs were rigidly flexed at a right angle to the abdomen, and the calves were flexed upon the thighs. The legs were strongly adducted so that the knees could be separated only with difficulty. There was no power of voluntary movement in either leg. The muscular rigidity was so great the legs could not be straightened. The rigidity was not caused by active muscular spasm. The right arm was also completely palsied and contractured in flexion at the elbow and wrist. The knee jerks could not be obtained, but this may have been on account of the extreme muscular rigidity. The muscle jerks were absent. The plantar jerk also was absent. There was no palsy of the left arm. He did not speak at all, paid no attention to anything said to him, and gave no evidence of intelligence. He would swallow food put in his mouth. Sensibility to touch and pain could not be determined because of his mental state. He died on January 15, 1905.

The necropsy revealed chronic myocarditis with disease of the coronary arteries, marked general arterial sclerosis, chronic interstitial nephritis, and areas of cerebral softening. On gross examination of the brain there was found an extensive area of softening involving a large part of the left hemisphere. The only parts that escaped destruction superficially were the first and second frontal convolutions and the anterior portion of the third, the apex of the ascending frontal and ascending parietal, the superior parietal, the third or inferior occipital, the lower part of the second and third temporal, and a small convolution situated in the centre of the softened area in about the position of the lowest part of the ascending parietal, which must have obtained its blood supply from a vessel coming from the interior of the brain at that point. The area of softening surrounded this spot and it is hardly possible that any fibres from it escaped destruction. The mesial surface of the left hemisphere of the cerebrum was normal. On the right hemisphere there was an area of softening involving the calcarine fissure practically in its whole length (about 3 cm.) and about 1 cm. in depth. It was entirely on the mesial surface, the lateral surface of the right hemisphere appearing normal. On the basal aspect the convolutions were normal. The arteries at the base of the brain were intensely sclerotic. On section the area of degeneration in the left hemisphere was seen to extend into and involve the anterior and part of the posterior limb of the internal capsule so that fibres from any part of the left cortex except the occipital lobe would be interrupted in some part of their course. The internal capsule on the right side appeared normal. On microscopic examination the right internal capsule was found normal by the Weigert hæmatoxylin and hæmalaun acid-fuchsin methods. The left internal capsule was largely involved in the lesion of the left hemisphere. There were a few normal fibres in the posterior part of the posterior limb, but most of them appeared degenerated by the Weigert method. The right cerebral peduncle was normal. No degeneration was seen in it by the Weigert method. Sections from the left cerebral peduncle showed the whole crusta distinctly smaller than the right and the nerve fibres of the pyramidal tract completely degenerated by the Weigert method. There was an area in the internal quarter of the crusta which was only partially degenerated. Turck's column was degenerated, showing that fibres from the second and third temporal convolutions must have been interrupted in some part of their course. Sections from the pons showed complete degeneration of the pyramidal tracts on the left side by the Weigert hæmatoxylin method. On the right side they were normal. In sections from the medulla the left pyramid was seen to be entirely degenerated by the Weigert method. The right pyramid and other parts of the medulla were normal by the Weigert and the hæmalaun acid-fuchsin method. Sections from the cervical region showed degeneration, by the Weigert method, in the left direct pyramidal tract and the right crossed pyramidal tract. The small intramedullary bloodvessels appeared very prominent in sections stained by the hæmalaun and acid-fuchsin stain because their walls were thickened. Sections from the midthoracic region were similar to those from the cervical region, the degeneration being confined to the left direct and right crossed pyramidal tracts. Sections from the second lumbar showed no other changes than above described. In the third lumbar the degeneration in the left direct pyramidal tract by the Weigert method was very slight, but distinct. In the right crossed pyramidal tract it was much the same as in the second lumbar. In the fifth lumbar there was distinct degeneration in the crossed pyramidal tract. The bloodvessels of the pia and also in the substance of the spinal cord and medulla showed marked arterial sclerotic changes, but no signs of inflammation or round-cell infiltration. The cells of the anterior horn of the spinal cord in the cervical region were normal by the thionin stain, but they were considerably pigmented and one or two showed slight granular changes in the chromaphilic elements. The cells in the anterior horn of the lumbar region, second, third, and fifth segments, were the same as in the cervical region. The optic chiasm and the optic nerves were normal by the hæmalaun acid-fuchsin and Weigert hæmatoxylin stains. In short, there was nothing in the brain or cord to explain the condition of the left leg. The cord showed only the degeneration proper to be found in the right-sided hemiplegia. The conditions under which the necropsy was made prevented an examination of the bloodvessels, nerves and muscles of the legs; so that it would be hazardous to draw any conclusions from this case alone. Fortunately we were enabled soon after to examine a case of paraplegia surely caused by arterial disease in the legs, and in which spinal-cord disease competent to produce the condition was absent. Since the arterial condition was the same in both cases, and since there was no disease of the brain or cord competent to explain the symptoms, and since pseudopalsy is well known to be caused by peripheral arterial disease, we think it explains the first as it surely does the second case.

The history of the second case is as follows:

G., a white man, aged sixty-eight years. He was senile and could give no accurate description of his illness on account of poor memory, which he himself realized. He lay in bed with the legs flexed on the thighs and the thighs on the abdomen. The muscular rigidity was very marked. He could flex and extend the thighs and calves a little and could lift the foot a short distance from the bed, but of course could not walk. Attempts at passive movements of either leg immediately increased the rigidity, which, however, could be overcome. There was a very slight occasional knee jerk on the right side, but none on the left. The muscle jerks were absent. He had incontinence of urine and feces. Some weeks before death sensibility was normal on the legs, but whether they became anæsthetic before death we do not know. There was no deformity of the spine nor pain on pressure over it. There was slight muscular wasting in both legs. Later he developed gangrene of the foot and died February 1, 1905.

Necropsy Report. The immediate cause of death was the rupture of an intercostal artery. There were also general arterial sclerosis of high grade, general senile atrophic changes in the abdominal viscera, cardiac hypertrophy, valvular sclerosis, and coronary atheroma.

The brain, spinal cord, and pieces of the posterior tibial artery, nerve, and muscle from each side were removed and preserved in Müller's fluid. The brain was well developed and there was no atrophy or areas of sclerosis. The convolutions were somewhat flattened over both hemispheres. The dura was normal. The pia was slightly thickened, but not adherent. The arteries of the brain, especially at the base, were very sclerotic. The lateral ventricles were quite dilated. The distance between the posterior horn of the lateral ventricles and the posterior pole of the brain was only one cm.; from the lateral wall of the posterior horn to the parietal surface was 2.5 cm., and from the anterior horn to the anterior pole was 3 cm. The ependymal lining of the ventricle, especially of the anterior horn, was distinctly roughened. The cortical gray matter was about normal in thickness. The foramen of Monro was distended and the choroid plexus cystic. The basal ganglia and internal capsule were normal. The aqueduct of Sylvius was patulous, the fourth ventricle about normal in size, and serial sections of the medulla showed no obstruction in the central canal below the fourth ventricle. The cerebellum, pons Varolii, medulla oblongata, and spinal cord appeared normal on gross examination.

The cortex of the paracentral lobules on each side was microscopically normal. The pia was slightly thickened. The Betz cells contained a large but (considering the patient's age) not abnormal amount of yellow pigment situated at one pole of the cell. The chromophilic elements generally stained well, a few cells only showing a granular appearance of the Nissl bodies. The bloodvessel walls were thickened, but there was no round-cell infiltration about them in the pia or cortex. Sections from the optic chiasm and from the optic nerves appeared normal when stained with the hæmalaun and acid-fuchsin and by the Wiegert hæmatoxylin methods. Sections from the medulla oblongata stained by the hæmalaun and acid-fuchsin methods apeared normal but for thickening of the walls of the bloodvessels both in the pia and medullary substance. The anterior pyramids were well stained by the Weigert hæmatoxylin method. Sections from the cervical region of the spinal cord stained by the hæmalaun acid-fuchsin method did not show the changes found in arterial sclerosis of the spinal cord—namely, an overgrowth of glia tissue about the periphery and about the bloodvessels of the cord, with a disappearance of nerve fibres. The bloodvessels in the pia and in the substance of the cord were distinctly thickened, but there was no overgrowth of glia about them. The pia was not thickened and there was no round-cell infiltration in it or about the bloodvessels in the cord.

The Weigert hæmatoxylin method showed distinct degeneration in the inner portion of Burdach's columns and in the columns of Goll, especially in the posterior portion. The Weigert method showed no degeneration of the posterior roots and entrance root zones. Sections from the cervical cord stained by the Marchi method showed recent degeneration in the entrance root zones and in the columns of Burdach. It was very slight in the columns of Goll. Fibres entering the cord showed degeneration by the Marchi method, but posterior roots attached to the section were normal. There was no recent degeneration in other parts of the section. Sections from the thoracic and lumbar region stained by the hæmalaun acid-fuchsin stains showed the same conditions present as described in the cervical region. Sections from the lower thoracic region stained by the Weigert method showed degeneration in the posterior columns more marked in Burdach's columns and the anterior portion of the columns of Goll. The entrance root zone did not show degeneration by the Weigert method. The Marchi method showed marked recent degeneration in the columns of Burdach and in the fibres of the posterior roots after passing through the pia. The posterior roots outside the pia showed no degeneration. Sections from the lumbar region showed a diffuse degeneration of the posterior columns by the Weigert hæmatoxylin method, which, however, did not extend as far forward as the commissure or as far back as the pia. The entrance root zone and the anterior and lateral columns were not degenerated. In sections from the lumbar region stained by the Marchi method the recent degeneration was seen beginning where the posterior roots pass through the pia and extending directly to the degenerated part of the posterior column, the middle third, on each side. There was no degeneration in other parts of the section. Posterior lumbar roots, cut separately from the cord, showed no degeneration by the Marchi method. The cells of the anterior horns of the spinal cord in the cervical, thoracic, and lumbar regions were studied with the thionin stain. They contained a very large amount of pigment. Occasionally the pigment occupied the entire cell and the nucleus was not visible. The chromaphilic substance was usually well stained. In one or two cells (from twenty-four sections examined) there was chromatolysis with displacement of the nucleus to the periphery. Considering the age of the patient, the anterior horn cells may be regarded as normal. The posterior tibial nerve from the right side was almost completely degenerated. The sheath of the nerve was very thick and there was great overgrowth of connective tissue between the bundles of fibres. The nerve fibres themselves in many of the bundles had been replaced by fibrous tissue. But few normal axis cylinders were seen. There appeared to be no inflammatory process or round-cell infiltration. Sections of the left posterior tibial nerve had the same appearance as the right. The right tibial artery showed such marked thickening of the wall that its lumen was almost closed. The left posterior tibial artery was closed by an organized thrombus. Sections from the right and left posterior tibial muscles showed great atrophy. The muscle fibres had a rounded appearance and were separated by an overgrowth of fibrous tissue. Small bloodvessels seen in the sections showed almost complete closure by the thickening of their walls. No nerve fibres were seen in the sections of muscles stained by the Weigert method. The intramuscular nerve filaments had probably degenerated completely. To sum up, there was moderate dilatation of the lateral ventricles, some degeneration of the posterior columns of the cord, great general arterial sclerosis so marked in the right posterior tibial artery as almost to occlude it, while the left was completely closed by an organized thrombus, muscular atrophy, and degeneration of the peripheral nerves. The changes in the spinal cord were not sufficient nor of a nature to have caused paralysis in the legs. The changes in the muscles and nerves surely resulted from the vascular disease. The case, then, was one of senile paraplegia caused by obliterating arteritis. This condition is not very infrequent. It may come on slowly with dull leg pains, paræsthesias, and gradual loss of muscular power, or if thrombi form the onset may be sudden and the result gangrene. In many cases in the earlier stages intermittent lameness occurs.

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A CASE OF TRAUMATIC CERVICAL HEMATOMYELIA AND COMPLETE DIVISION OF THE CORD, WITH PROBABLE DISLOCATION OF THE FIFTH CERVICAL VERTEBRA. REMARKS ON THE LOCATION OF THE CENTER FOR THE BICEPS REFLEX.[1]

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Summary.—A white male aged 56 years, who after a fall down stairs was immediately rendered unconscious for about twelve hours. On recovering from which, with the exception of some delirium several days later, mentality was good. Complete motor and sensory paralysis involving the legs and trunk and partially involving the arms. Loss of all tendon reflexes excepting the biceps, which, however, was lost temporarily on the right side. Preservation of the cilio-spinal, cremasteric and plantar reflexes, the latter disappearing shorting before death. No Babinski phenomenon. Death on the ninth day. At the autopsy was found a possible slight displacement of the fifth cervical vetebra; marked compression of the cord, so as to practically divide it between the sixth and seventh cervical segments. Hemorrhage into the gray matter extending from the eighth cervical up to and partially destroying the right posterior horn of the fifth segment. There was also a fracture of the right side of the skull, with extensive extradural hemorrhage compressing the motor area, which, excepting the early period of unconsciousness, had caused no symptoms.

History.—A. C., aged 56 years, was admitted to the Nervous Wards of the Philadelphia General Hospital on September 11, 1904. His temperature being 101 degrees, pulse 90, and respiration 35 to the minute. He stated that about twenty-four hours before admission, after drinking several glasses of ale, he had fallen down stairs, and was immediately rendered unconscious, remaining so for about twelve hours. When he recovered consciousness it was found that the legs and arms were paralyzed. Examination September 12, about 36 hours after the receipt of the injury, showed a well-developed man, who answered questions intelligently and stated nothing of note concerning his previous or family history. The temperature, pulse and respiration were normal. He lay in bed on his back with the arms elevated about 15 degrees from the body, the forearms flexed on the arm and somewhat supinated, and the fingers flexed.

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[1] From the Neuropathological Laboratory of the University of Pennsylvania and from the Philadelphia General Hospital. Read before the Philadelphia Neurological Society, Dec. 27, 1904.

The pupils were equal and responded to light and accommodation. There was no paralysis of any of the cranial nerves. He could elevate each arm to a right angle with the trunk, and they could be rotated. Flexion of both forearms could be performed, but not with normal strength, the forearms could be slightly supinated. The abdomen was somewhat distended. The legs were completely paralyzed. No atrophy was noticed. Electrical reactions were not taken. The muscles were flaccid.

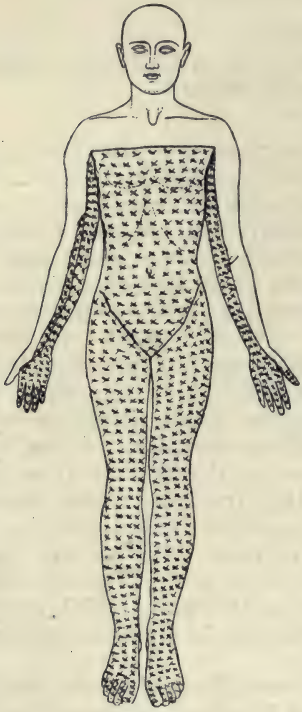

FIG. 1.

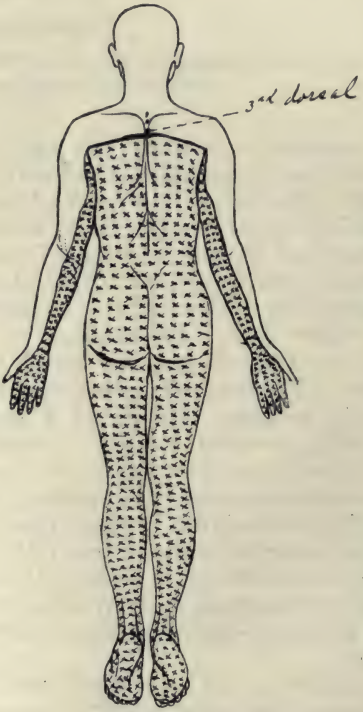

FIG. 2.

The cilio-spinal reflex was present on both sides, the biceps jerk was present and active on both sides, the triceps, wrist, knee and Achilles tendon jerks were absent, there was an active cremaster reflex, and when the soles of the feet were stroked there was slight flexion of the toes. Retention of urine and constipation were present and at times there was slight priapism.

The distribution of the sensory paralysis (tactile, pain and temperature) was as is indicated in Figs. 1 and 2.

The patient complained of pain in the back of the neck, and there was some tenderness in the lower cervical region, but no deformity was visible, either to the eye or in the skiagraph. Bed sores were beginning over the right trochanter and the scapulæ, and the stick of a pin was followed by an area of hyperemia which persisted for some time.

The urine was acid and contained albumin and hyaline and granular casts.

On September 14 it was noted that the patient was delirious, but could be made to answer questions and obey commands. The abdomen was distended and tympanitic. During respiration the abdominal muscles did not contract for several seconds; they then contracted spasmodically one or more times, to be followed by a period of rest as before. The arms could be elevated, and rotated outward further than at the first examination, otherwise there was no change. On September 16 it was found that the toes were neither flexed nor extended when the soles of the feet were irritated. On the right side the biceps jerk was absent, but was very active on the left side. He was delirious at times but answered questions intelligently when spoken to. The peculiar respiratory movements noted on the 14th had disappeared. The left pupil was slightly dilated, but both responded to light and in accommodation. The cilio-spinal reflex could not be obtained. The temperature was 96, pulse 40; on the 17th these again became normal; respiration normal. On Sept. 18 Dr. Weisenburg noted that the patient's face was cyanotic, the left pupil dilated, and the left palpebral fissure slightly narrowed. His voice was weaker, and he could not cough as well, but he could swallow. A slight biceps jerk on the right side was obtained. The patient died on September 19, 1904. The autopsy was made about twenty-four hours after death by Dr. A. G. Ellis, Dr. Weisenburg being present, to whom I am indebted for the following report:—

The scalp in the left frontal and the left parieto-occipital region is ecchymosed. There is a fracture of the skull, starting from a point one-half inch to the right of the median line, at the junction of the parietal with the occipital bone. The line of the fracture is to the left, forward and downward, and ends in the squamous portion of the temporal bone just in front of the ear.

The skull is unusually and uniformly thin, at its thickest portion not measuring more than one-eighth of an inch. Inside of the skull, in the distribution of the left middle meningeal artery there is an extensive extradural hemorrhage. This lies behind the central fissure. The brain, when removed, shows a depression in the region of the hemorrhage, but otherwise nothing is found macroscopically.

The tissues surrounding the upper cervical vertebræ are congested. There is apparently no fracture or dislocation of the cervical vertebræ[2], at least the vertebræ are not movable and crepitation is not felt. The cervical vertebræ from the seventh to the third inclusive, were removed intact, and show nothing abnormal, except that opposite the fourth and fifth cervical vertebræ there are adherent clots of blood. The upper portion of the third cervical and the second cervical vertebræ were removed piece-meal.

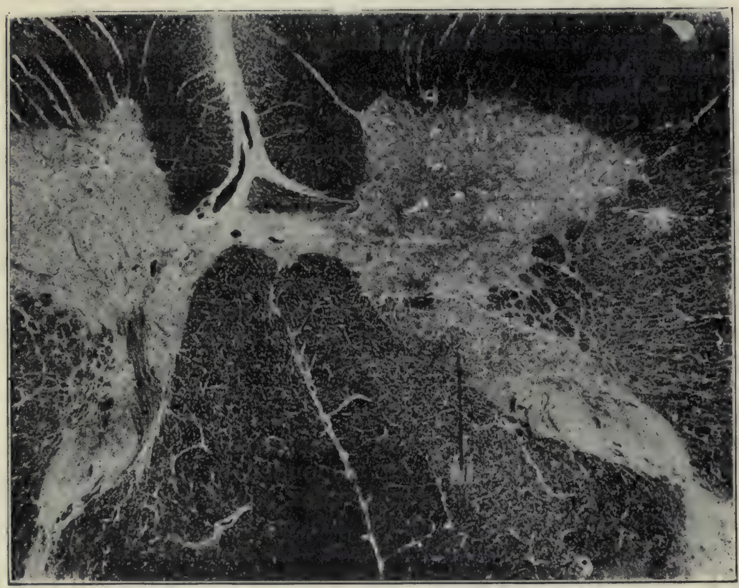

FIG. 3.—Photograph of a section from the fifth cervical segment showing an area of softening and hemorrhage in the right posterior horn (A).

The spinal cord in its upper portion, between the sixth and seventh cervical segments, showed a transverse depression, the cord measuring here about one-eighth of an inch. For an inch both above and below this part the cord was soft to pressure.

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[2] Dr. Ellis thought that the fourth cervical vertebra was displaced slightly backward, but from the location of the compressed area of the cord it is more likely that the fifth had been displaced and then was spontaneously reduced.

Microscopical Examination.—The compression, which is very intense, is between the sixth and seventh cervical segments. Sections were made through the lower part of the fifth cervical segment, which was the utmost limit of the area of softening. The area of compression was not cut, as it was unnecessary and it was not thought advisable to destroy uselessly a valuable gross specimen.

At the level of the second and third cervical segments the spinal cord shows no alteration. There is no ascending or descending degeneration even by the Marchi method, as the patient only lived one week after the accident.

At the level of the fifth cervical segment the right posterior horn is destroyed in its ventral half by softening and hemorrhage. (Fig. 3) The posterior half appears to be normal. Fibers are seen entering the right posterior horn and stopping a little short of the area of softening within the horn. The softening seems to be almost confined to the right posterior horn. This is the only portion of the cord at this level which seems to be pathologically altered, the left posterior horn apparently being normal. It is presumable from a study of the sections that the posterior root supply of the fifth cervical segment must have been impaired on the right side.

Sections taken from the lower part of the fifth cervical segment stained by thionin show considerable alteration of many of the nerve cells. In a number of these the nucleus is displaced to the periphery and the chromophilic elements are disintegrated and appear as fine granules, and some of the cells are swollen. This alteration is seen in the anterior horn of each side.

In the lower part of the eighth cervical or the upper part of the first thoracic segment the cells of the anterior horn are very little diseased.

The spinal cord at the first thoracic segment and below appears to be normal. The twelfth thoracic and the second and fourth lumbar segments are perfectly normal. Special attention has been directed to this part of the cord, because of the loss of all the tendon reflexes.

The most interesting feature of this case is the evidence that it affords of the location of the center for the biceps jerk. This is usually given as the fifth and sixth segments. On the left side in this case the biceps jerk was very active until the death of the patient, while on the right it was active for the first couple of days, then disappeared, to again become feebly present two days later. Microscopic examination of the specimen shows that on the left side the fifth segment is intact, while on the right side the posterior horn is partially destroyed by hemorrhage and softening. (Fig. 3). It seems probable therefore that at first this segment was not involved on the right side, but later became so by the hemorrhage gradually extending upward, at which time the reflex disappeared; then as some absorption took place, the functions of the segment partially returned, and it again became feebly present. As the biceps jerk was very active while the fifth segment was intact, it seems likely that this segment has most to do with the biceps jerk, and that the sixth plays only a subordinate part, if any.

The fact that the eighth cervical and first dorsal segments were intact accounts for the preservation of the cilio-spinal reflex.

While Bastian claimed that both superficial and deep reflexes are abolished in complete transsection of the cord, this case conforms to what has usually been noted by observers, i.e., the cremaster and plantar reflexes were present, it differs however in the fact that the plantar reflex gave the flexor and not the extensor response, which according to Turner is the rule. This possibly might have developed if the patient had lived longer. Attention is called to the fact that there was no lesion in the lumbar cord.

The distribution of the motor paralysis corresponds to the muscles usually regarded as supplied by the affected segments.

The area of sensory paralysis does not correspond to the diagrams of Head, but with the exception of the non-involvement of the thumb is similar to those of Starr and Kocher.

That there was an extensive extradural hemorrhage over the motor area of the left side which gave no symptoms, if we except the period of unconsciousness present immediately after the fall and a possible weakness of the muscles about the angle of the mouth in the right side when the man was admitted, but which had disappeared the next day, is a fact of interest.

NOTE—Since the foregoing was written Dr. Morton Prince has reported in the JOURNAL OF NERVOUS AND MENTAL DISEASE, Feb., 1903, p. 81, a case in which one side of the cord was severed between the sixth and seventh cervical segments. The area of anæsthesia in his case is similar to that in the above case, i.e., the thumb is not involved, and in this regard differs from the diagrams of Starr, Kocher and Wichman.

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ENCEPHALITIS AND OTHER NERVOUS AFFECTIONS COMPLICATING SCARLATINA.*

BY

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The following case is important and interesting chiefly because while clinicly, symptoms of meningitis or irritation of the brain were practically absent, pathologicly there was evidence of beginning pachymeningitis and encephalitis.

The patient, a girl of 9, was admitted to the Municipal Hospital on December 30, 1904. The family history was negative, and the previous history was unobtainable beyond the fact that she had had measles just prior to her admission. The eruption of scarlet fever appeared just before she was admitted, having been preceded by vomiting, fever, and angina. There was nothing especially interesting in the course of the disease, except that nephritis intervened, and, a short time before death, there developed a tremulous condition of the general musculature. The temperature ranged between 100° and 106°, and the pulse varied from 90 to 160. Upon admission there was a mild delirium, which disappeared in a day or two. She was comatose a few hours before death.

The autopsy, performed by F. Burvill Holmes, established a pathologic diagnosis of edema of the lungs, fibrinous pericarditis, acute splenitis, acute endocolitis, and subacute parenchymatous nephritis. The brain was hardened in formalin, and later, portions were placed in Müller's fluid. Sections were made from various portions of the brain, and stained by the Weigert hematoxylin and Cajal neurofibril methods, by thionin, hemalum, and acid fuchsin. Studies were also made with the Löffler stain.

In the right paracentral region there was a distinct, though not intense round-cell infiltration of the pia, and the pia was slightly thickened. One subpial hemorrhage was seen. Within the substance of the brain there was a moderate perivascular round-cell infiltration. The bloodvessels were congested, and occasionally a leakage of blood out into the tissue was observed. In the sections stained by thionin no change could be observed, except that the typhoid substance of some of the nerve cells was somewhat granular.

In the frontal region there was distinct distention of the perivascular spaces, and a marked congestion of the bloodvessels. Round-cell infiltration of the pia and small subpial hemorrhages were present also in this part of the brain. In the temporal region the vessels were very prominent, and here also there was some distention of the perivascular spaces.

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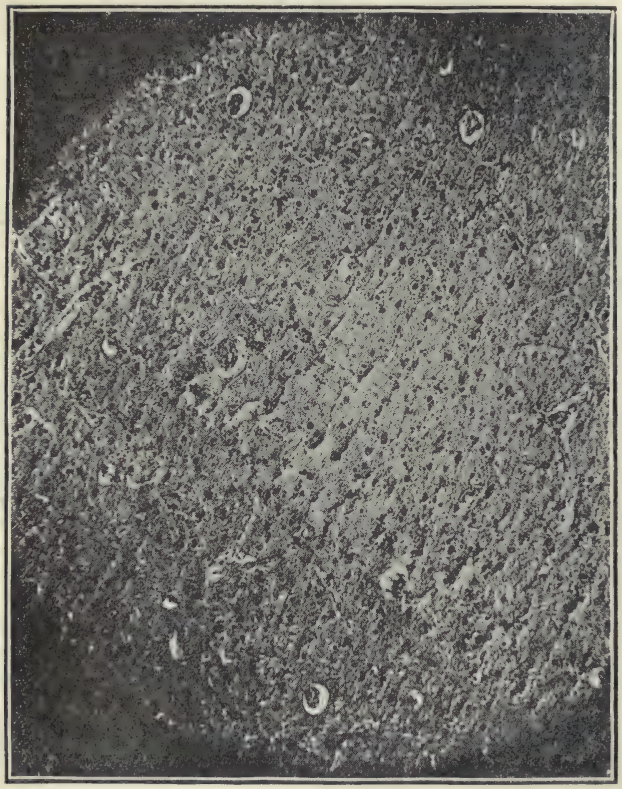
* Read before the Philadelphia Pediatric Society, May 9, 1905.

In the occipital region there were several microscopic hemorrhages into the substance of the brain and under the pia. The pia was thickened, and there was a moderate amount of round-cell infiltration. The pons and cord were examined, but nothing pathologic was found. Sections from the paracentral region, stained by the Löffler stain, failed to show the presence of any microorganisms.



The presence of a mild meningitis associated with an undoubted, if moderate, inflammatory change in the cortex of the brain occurring as a complication of scarlatina, is a finding which, to say the least, is unusual, possibly because it is not frequently searched for in such cases. While meningitis has been observed in a number of cases of this disease, little is known about the part that scarlatina plays in the production of encephalitis, and with the exception of the statement made by Stiebel,[61] that the exanthems bear very little relation to the production of encephalitis, I have found no reference in the literature to its occurrence in scarlatina except the recent report of Southard and Sims. These authors,[63] in September, 1904, reported a case in a boy of 5, who developed in the fifth week of an attack of scarlet fever a slight limp associated with violent contractures of the flexors of the right thigh. Later all the muscles of the right side, with the exception of those of the face, were attacked with regular contractions and relaxations. The right arm was paralyzed, and pain and tactile senses were rather impaired. Complete paralysis of the right side followed a subsequent attack of contractions. The right side of the face was also involved, and the patient died at the end of two weeks. The pia mater and arachnoid were the seat of a purulent infiltration. Numerous hemorrhages were found in the cortex, and phagocytic cells were found in the meshes of the pia and adventitia of the short arteries of the cortex. Phagocytosis and fatty change of the neighboring nerve tissues were observed, as well as an axonal degeneration in the cells of the anterior horns of the cord. Southard and Keene[64] published later the same case with five others presenting different clinical symptoms but in which, postmortem, *Staphylococcus pyogenes aureus* was found in the brain. They conclude from their study of these cases, together with the results of experiments on animals, (1) that *Staphylococcus pyogenes aureus* produces in the meninges and brain substance of man a type of inflammation in which hemorrhage is prominent (subcortical region), and a diapedesis, and slight leukocytic emigration up to abscess formation, and acutely destructive hemorrhages with phagocytosis are present. The acute inflammatory appearance was often obscured by collections of mononuclear cells, which were phagocytic for cells and cell detritus; (2) that *Staphylococcus pyogenes aureus* produces an inflammatory process in the brains of guinea-pigs, which tends to subside within two weeks without clinical symptoms. The cell picture is of meningitis at the end of 6 hours, of ependymitis in 12 to 24 hours, and exudation in 24 to 48 hours; (3) that the lesion in pigs is curable; in man irreparable. This is a very important contribution to the pathology of this subject.

The findings in my own case were in some respects similar to those found by these investigators. Numerous small hemorrhages were seen in the brain cortex, and effusion of blood under the pia was present in several places, but the changes in my case were conspicuously inflammatory, though the process had not gone very far and the staphylococcus was not found.

In June, 1904, I[54] described a somewhat similar lesion occurring in a child who, after pertussis, developed spastic diplegia. In the brain substance there were found small hemorrhages, and greatly distended perivascular spaces partially filled with round cells. There were also acute degeneration of the brain tissues and a round-cell infiltration of the meninges. During the folollowing summer Neurath described somewhat similar changes in the brains in a number of fatal cases of pertussis.

I think we may conclude that meningitis and encephalitis of an inflammatory type, occur as a result of the irritation set up by the streptococcus, or of a toxin in the blood emanating from this, or from other organisms. Strümpell, in 1884, advanced the theory that in acute infections, encephalitis, analogous to acute myelitis in the anterior horns of the cord, was responsible for the development of infantile hemiplegia. Landon Carter Gray[31] viewed this theory as a "flippant" pathologic suggestion, and Gowers stated that a primary inflammation of the cortex of the brain is a purely theoretic disease. Marie states that primary acute myelitis does not exist, but is always the result of an infection.

In the light of the knowledge thrown upon the subject by the cases of Southard and Keene, Neurath's[50] and my own case, more weight must be given to Strümpell's theory, and just as acute myelitis is caused by an infection irritant, so may we see acute encephalitis arise from a similar cause.

How often idiocy—a frequent sequel of scarlatina—and epilepsy may be caused by this very process is of course speculative, but that either may be a result is not untenable.

Extension from neighboring inflammations, such as middle-ear disease, or from infectious embolism may produce encephalitis, as Huguenin[33] and others have shown. In Tonkin's[66] case a purulent focus was found in the left frontal region, causing death.

Meningitis itself may be a complication of scarlatina, though, according to Eichhorst[23] and Barthez and Sauné,[4] it is a rare one. If microscopic examinations of the brain in cases of scarlatina were less uncommon, probably meningitis would be found more frequently in these cases. Claussnitzer[19] in an inaugural dissertation entitled Ueber hæmorrhagische Meningitis bei Scharlach, declares that while primary meningitis exists in this disease, it must be looked upon as of infrequent occurrence, and that in the majority of cases exhibiting meningeal symptoms, which have come to autopsy, there is merely hyperemia of the brain and meninges. He found only 17 cases in the literature, of genuine inflammation of the meninges. He cites 2 cases of his own, one in a girl of 18, exhibiting symptoms of meningitis, the autopsy revealing the presence of an inflammatory change in the membranes; and a second case in which, without signs of meningeal irritation, there was found at the autopsy hyperemia of the meninges and brain substance, beside small hemorrhages between the pia and cortex, and round-cell accumulation.

Stiebel,[61] as early as 1859, recognized that "arachnoiditis" and more rarely, "inflammation of the pia" complicated scarlatina.

Litten[42] stated that the brain was never affected in his uncomplicated cases, although he recognized the existence of pachymeningitis hæmorrhagica, as well as inflammation of the meninges in these cases. Other cases have been recorded by Volz, Thomas, Halbey (cited by Claussnitzer) Mayr and Fischer (cited by Barthez and Sauné).

Bendel[3] cited a case of meningitis in a child of 4, exhibiting meningeal symptoms, namely, hyperesthesia of the skin, the hydrocephalic cry, right-sided hemiplegia, convulsions and death. There was no autopsy. A second case occurred in a girl of 12, who suffered from coma, trismus, spasm of the neck muscles, and clonic and tonic convulsions of the muscles; death resulted. A sister died exhibiting similar symptoms. Two other somewhat similar cases are cited. There was no autopsy made in these cases.

The commonest complication of this exanthem is hemiplegia. Bernhardt[5] reported 4 cases of this character, 2 with and 2 without aphasia. Willems[73] collected 65 cases of hemiplegia in children, of which number 5 followed this disease. He reviewed the literature of the condition rather carefully, and quoted a case of Delius, reported as early as 1777, and one of Suedois-Hagström, reported in 1790. He reported a case of his own in a woman of 28, who developed, on the eighth day of the attack, pain in, and paralysis of, the arms. All symptoms disappeared in three weeks. Willems believed this was of toxic origin.

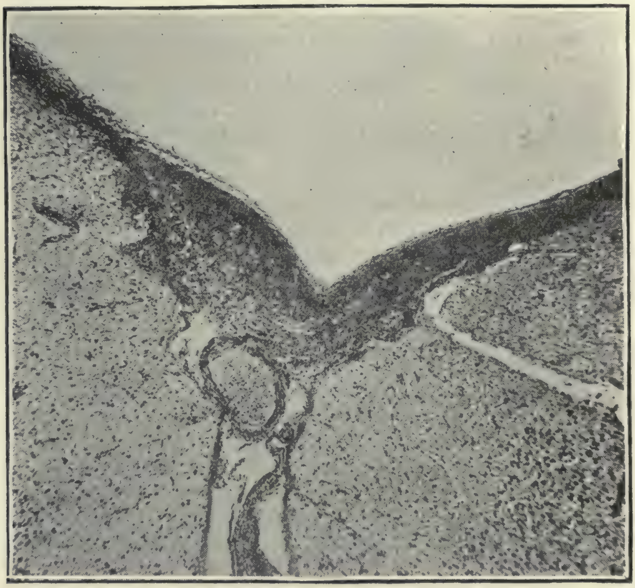
In two cases of hemiplegia reported by Terrier,[68] there were incontinence of urine and feces and epileptiform convulsions. The reflexes were increased, and Babinski's phenomenon was present. In Barlow's case, a girl of 7, there suddenly developed right hemiplegia two months after a mild attack of scarlet fever. Barlow[13] concluded it was due to cardiac thrombosis, the result of an embolism.

In a case reported by Henoch[34] a child of 3 developed a sudden hemiplegia in the fifth day of an attack of scarlet fever. The patient recovered entirely in a year. Sachs,[65] in a study of 83 cases of hemiplegia in children, found scarlatina the cause in 3 cases, while Osler[51] attributed the cause to infectious disease in 19 out of 135 cases. John T. Walker[74] cited a case of scarlet fever in a girl of 7, ushered in by convulsions lasting six hours, while fever, followed by coma and hemiplegia, developed later. Great improvement in the paralysis followed, and while the gait was imperfect, the patient was able to walk, though not to run. Similar cases have been cited by Pepper,[52] Condie and others.

Aphasia was associated with hemiplegia in a few cases reported by Eulenburg,[22] Bohn,[7] Bernhardt[5] and Addy.[1] In Bernhardt and Sachs' experience aphasia may accompany right or left hemiplegia in children. Aphasia has also occurred unassociated with hemiplegia. In Brasch's case, a girl of 3 years and 10 months, aphasia developed at the end of the first week. The patient recovered entirely.

In a case reported by Brill, an irregular aphasia followed several convulsive attacks, but improvement ensued after the administration of nitroglycerin. In a case reported by Sheppard,[59] a girl of 5 lost the power of speech during the first day of the disease. A month later she was generally palsied, and some three weeks later she was able to walk, but her gait was distinctly ataxic. Great improvement followed. In Taylor's[70] case a boy of 5 became paralyzed in the second week of the fever. Some time later he died of diphtheria, and the necropsy revealed the presence of an embolism of the left middle cerebral artery, an area of softening in the posterior part of the second and third frontal convolutions, in the parietal and supramarginal convolutions, the island of Reil, the corpus striatum, and the anterior portion of the internal capsule. Vegetations were found on the valves of the heart.

The cause of the hemiplegia in these cases was either hemorrhage, embolism, or thrombosis. While there is no doubt that in most of the cases these are the causes, the case of Southard and Keene demonstrates the possibility of still another pathologic basis for its development.



The next most frequent complication is involvement of the peripheral nerves. A case of multiple neuritis is cited by Greene,[27] as occurring in an epileptic boy of 17, who had during an attack of scarlatina, 1,742 fits. He had practically recovered from the fever when the multiple neuritis developed. He recovered entirely in seven weeks.

Méry and Halle[47] describe the case of a boy of 7, who developed peripheral neuritis during an attack of scarlet fever. At the decline of the disease he suffered from two attacks of scarlatinal rheumatism. The authors ask whether this was due to a toxin the result of the scarlet fever, or whether it was a rheumatoid affection.

In a case reported by Sano,[60] a man of 42 developed paralysis of the right arm and shoulder on the seventh day of the disease. There were severe pain and contractures, especially in the biceps. The paralysis disappeared at the end of eight weeks, although some pain and atrophy remained in the muscles of the shoulder and arms.

Wood reported a somewhat similar case of paralysis of the right arm which afterward entirely disappeared. In the case of Egies, polyneuritis followed scarlet fever in a girl of 4. The peroneal nerve was palsied, and there was ataxia of all the extremities. The electric irritability of the ulnar and peripheral nerves was lowered, and there was tenderness along the nerve trunks. Cure was effected in two and a half months. Cases of peripheral neuritis are also cited by Thomas[72] and Moureyre.[45]

Thomas Barr[6] describes an interesting case of facial palsy in a girl of 4, which probably had some relation to suppuration of the middle-ear. Steffen[62] describes also a case of right facial palsy. In Ballard's[11] case, a boy of 2, there was paralysis of the laryngeal muscles five weeks after desquamation was completed. Paralysis of accommodation was present in a case cited by Malone.

Tetany has occurred in a few cases complicating this exanthem. Schatten[57] reported a case in a boy of 8 who, on the sixteenth day, developed tonic contractions of the muscles of three days' duration, associated with paresthesia, without other sensory disturbance or changes in the reflexes, or electric reactions. Steffen[62] also reported a case, and Kühn[38] refers to a case in a boy of 4½, who developed on the fifth day of the disease, stiffness and difficulty in moving the legs, which later extended to the entire body, lasting six weeks. He looked upon it as the result of a toxin.

In a case reported by McConnell,[46] tetany occurred during convalescence in a boy of 5½, resulting finally in entire recovery. Loeb[43] published the case of a girl of 6, who, after an attack of unconsciousness became tetanic in her thumbs and arms. There was increased irritability of both facial nerves. As the tetany improved, aphasia, choreiform movements, and ataxia followed, from which she finally recovered entirely.

Scarlet fever may cause choked disc, orbital optic neuritis, or amaurosis. In the case reported by Thomas,[67] a boy of 5 developed a short time after an attack of scarlet fever, progressive failure of sight caused by double optic neuritis. This was followed by paretic symptoms affecting all the extremities. The reflexes were exaggerated, and there was pronounced anesthesia. Speech was lost and the sphincters were involved. Four months later the patient had recovered largely, with some weakness in the right arm, associated with atrophy of the extensors and of the optic discs. In Thomas' opinion these symptoms were due to meningitis caused by the scarlatinal poison.

Amaurosis is probably of uremic origin, and the prognosis is good, the symptoms lasting usually only a few days. Reimer[56] and Loeb[43] have cited cases of this character, and the condition was described as early as 1769 by Withering (quoted by Imbert-Gourbeyre).[36] Gubler[30] refers to the case of Cazanave, a girl of 9 who gradually became blind during convalescence from an attack of scarlet fever. Later the sensations of taste and smell were lost.

A few cases are on record exhibiting ataxic symptoms. In the case of Greenhow,[24] a man of 40 suffered from numbness and tingling of the hands with loss of power, beginning at the end of the sixth week. Ataxic movements of the legs developed later, but all the symptoms disappeared in a few weeks.

Bruns[9] recorded a case exhibiting ataxic symptoms, in a girl of 7. In Sheppard's[59] case, a girl of 5 became generally palsied in the fourth week. Three weeks later she walked like an ataxic, but great improvement followed. Moureyre[45] described a somewhat similar case.

Eulenburg mentioned the fact that unilateral progressive atrophy of the face has followed scarlet fever in a few cases.

Epilepsy was described as a sequel of scarlet fever by Marie,[44] who cites the case of Landmann, in which petit mal followed scarlet fever in a girl of 12. At the age of 18 major attacks made their appearance. Other writers recognize it as a sequel also.

Disseminated sclerosis followed scarlet fever in a case of Madam Rosa Landis, cited by Raymond.[53]

Hysteria occurred in the case reported by Talairach,[21] and in one reported by Carnell.[15] In Carnell's case, a girl of 15, during convalescence from an attack of scarlet fever suffered from general weakness, which soon developed into paralysis of the arms, later involving the legs as well. The history states that the patient improved a great deal, and a diagnosis of hysteria was made.

A case of chronic hydrocephalus, following scarlatina, was reported by Demarge.[20] The autopsy revealed granulation of the ependyma of the ventricles, such as those described by Virchow under the name of papilloma, the result of chronic inflammation of the membranes of the ventricle.

Cases of paraplegia have been described by Althaus[2] and Kennedy.[40] In the case of Kennedy the patient recovered entirely, but in Althaus' case the injury was permanent.

Neuralgic affections are sometimes met as sequels to scarlatina. Gueneau de Mussy[25] (in Barthez et Sauné) refers to a case of facial neuralgia and in double facial neuralgia in one case, and sciatica in three cases observed by Roger.[55]

A case of Friedreich's ataxia following scarlatina is reported by Katz.[39]

Finally, mental diseases must be mentioned as complicating scarlet fever. Clouston[16] claims that it is the most frequent cause of postfebrile insanity, and Moureyre, who made a careful study of this subject, believed that scarlatinal psychoses are of very frequent occurrence. They are, he claimed, the result of the action of the scarlatinal poison upon the brain. The psychoses of the *periode d'état* may be (1) delirium of collapse, or mental confusion; (2) simple symptomatic mental confusion, in which there exists an acute hallucinatory confusion; and (3) the *psychoses de rêve.* These are very frequent. He cites 13 cases illustrating this classification.

Imbecility follows in the wake of scarlet fever; in fact, this disease was the cause attributed in 2.75% of the cases studied by Martin Barr,[8] and in 2.4% of Roger's cases (cited by Barr).

Very little is known about the lesions which cause paralysis and the other nervous phenomena following scarlatina, because the patients are very apt to return to health, or do not die at the climax of the nervous symptoms. Barthez and Sauné[4] state that in patients dying of convulsions the only alterations found are injections of the pia and brain. Meningitis has been found in a few cases and infiltration of the meninges, edema of the brain, cerebral anemia, congestion of the membranes, and subarachnoid ecchymoses represent the usual postmortem appearances. In the case of Tardieu[69] (in Barthez et Sauné) there was a cerebral hemorrhage the size of a pigeon's egg, and serous infiltration of the subarachnoid tissue.

Guinon[28] states that congestion of the pia and brain, distention of the cranial sinuses, and sometimes cerebral edema may be found, and rarely, thrombosis of the meningeal veins. According to Henoch,[34] the cerebral symptoms in scarlatina do not depend upon meningitis, but the postmortem changes consist of hyperemia, edema of the pia and brain substance, thrombosis of the sinuses, as well as stagnation, in consequence of lowered heart's action. Gerhardt,[29] in discussing this subject, stated that meningitis and purulent foci in the brain may be found. Thomas[72] put down as the anatomic findings, hyperemia, slight inflammations, edema, anemia, and hydrocephalus, with rarely severe abscesses and extravasations. Meningitis and effusions into the ventricles are uncommon, while apoplexy and sinus thrombosis are exceedingly rare. Very few cases in which the brain has been studied histologically have come to autopsy, and I know of only the case of Southard and Keene, and that of my own, in which a careful study of the brain substance, as well as the membranes, was made.

CONCLUSIONS.

The most common complications of scarlatina on the part of the nervous system are hemiplegia and peripheral neuritis. More rarely this disease may be followed by paraplegia, optic neuritis, amaurosis, tetany, pseudoataxia, neuralgia, epilepsy, disseminated sclerosis, Friedreich's ataxia, hysteria, chorea, hydrocephalus, meningitis, and disordered mental states.

With the exception of hemiplegia, and imbecility, the prognosis is good, if we exclude those rare organic cases such as Friedreich's ataxia, disseminated sclerosis, and epilepsy, which are recorded as following scarlatina. Some of these conditions like Friedreich's ataxia are probably merely hastened in their manifestations by the scarlatinal process.

The pathologic findings consist of thrombosis, embolism, small cerebral hemorrhage, rarely abscess of the brain, congestion of the brain, and meningitis, and finally meningitis and encephalitis.

I am indebted to B. F. Royer for the specimen and notes of the case, and to Robert D. Rhein for assistance in obtaining the references.

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A CASE OF VISUAL HALLUCINATIONS AND CROSSED AMBLYOPIA WITH VASCULAR AND DEGENERATIVE LESIONS IN THE CALCARINE CORTEX AND OTHER PORTIONS OF THE OCCIPITAL LOBE; ALSO WITH ATROPHY OF THE PREGENICULÆ AND OPTIC TRACTS.[1]

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The case briefly recorded below is of much interest from several points of view. It throws some light upon the question of the organic basis of visual hallucinations, and affords some valuable data regarding the pathological anatomy of the insanity of arterio capillary sclerosis. It also furnishes an illustration of a somewhat irregular form of crossed amblyopia, apparently due to lesion of the occipital cortex and subcortex.

The patient, a woman sixty-three years old, had suffered with impairment of vision in both eyes for nearly two years before coming under observation. This dimness of sight was attributed to glaucoma, for which a double iridectomy was performed. After this operation her vision was good in the left eye, but only fair in the right eye. She was not, however, blind in either eye and was able to do considerable near work, although she was careful in this respect. The following is a brief record of the state of her vision and of her fields shortly after the iridectomies, for which we are indebted to Dr. Geo. C. Harlan under whose care she was at that time and by whom the operations were performed.

At this time the vision in the right eye was 20/40 and the field practically normal with a slight peripheral cut in the temporal side; the disc was cupped, 1.50 D; vision in the left eye was 20/50; the field and the ophthalmoscopic examinations were the same as in the other eye.

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[1] Read at a meeting of the American Medico-Psychological Association, San Antonio, Texas, April 18-21, 1905.

In September, 1904, one of us (Dr. Mills) was called to Virginia to see this patient. A few days before she was seen she had had a cerebral seizure in which the right eye became totally blind and vision in the left eye was greatly impaired. Examination showed this complete loss of vision in the right eye, and so great a contraction of the fields of the left eye as to give the case the appearance of one of so-called barrel vision. At the time of her attack she lost the power of coordinating her movements sufficiently to stand and she was slightly delirious. She was mildly excitable, talking rapidly and somewhat inconsequentially. The members of her family regarded her as simply excited over her loss of sight and inability to stand, but her condition was one suggestive of hypomania. From the first it seemed that active mental disorder was imminent.

After a few days she was brought to Philadelphia where she was attended until her death by Dr. Harlan and Dr. Mills. Shortly after her arrival Dr. Harlan made a careful examination of her eyes. In the right eye she had scarcely more than light perception. The fundus was the same as in the previously reported examination, except that the cupped nerve had filled up to the level of the retina which equalled a swelling of plus 1.50; tension was normal. In the left eye vision was reduced to 5/40. The field was peripherally contracted to 40 degrees on the mesal side and to 20 degrees on the temporal side. The color proportion of the field was normal. The disc was slightly cupped, the fundus was otherwise normal.

On account of the patient's nervous condition it was not considered wise to make a minute outline of the fields.

Early in the history of this case there was something hard to describe in her mental state and attitude. She talked and acted as one somewhat emotionally exalted and lacking in inhibition. As she had not at first any hallucinations or illusions, some of those around her were inclined to regard her as not suffering from any mental unbalance, she normally being vivacious and inclined, on slight cause, to excitement. At first she was not able to stand or walk, but in the course of ten days she became able to walk without support for a few feet. How far her inability was mental and how far physical, it was difficult to say, as under strong encouragement she stood and walked better.

Two weeks after arriving in Philadelphia she suddenly became violently delirious during the night, talking and screaming and showing signs of visual hallucinations. She leaped from the bed and became difficult to manage. From this time on until a few days before her death, when she became quieter through weakness, she was acutely maniacal.

A record of a few of her hallucinations will be of interest. Four days after she had given clear evidences of the existence of acute mania, she saw men in the room with knives and pistols assaulting her son and husband as well as herself; she said that her own life was in danger but she did not care for that if only her son and husband were saved. At about this time she also saw fire occasionally, but she still continued to recognize those around her, and talked with them more or less rationally. She also now and then spoke of some dead relatives. Up to this period she had shown no fever, although her temperature had occasionally arisen to 99 or even to 100 degrees.

About one month after coming under continuous observation she developed a glandular swelling on the under right side of the face which disappeared in a few days. She died about three months after coming under care in Philadelphia. During most of this time illusions and hallucinations of sight were the most marked features of the case. She would, for example, suddenly cry out that she saw persons or objects threatening her or passing before her. She sometimes mistook the nurse or the doctor for someone with evil designs against her. With an effort her attention could be temporarily fixed, but she would suddenly cry out with great fear, pointing in the direction of some imaginary person or object. The hallucinations and illusions of sight kept up until a state of thorough exhaustion came on a few days before her death. They were not associated with hallucinations of sound, and there was no involvement of other senses unless the statements which she made at one time early after the development of visual hallucinations, that her legs were being or had been cut off, might be so considered.

Examinations of her urine were frequently made and on one or two occasions showed a few casts and on one or two others a trace of albumin; otherwise there was nothing of special importance. The blood examination made about ten weeks before her death showed: red cells 4,280,000; white cells 8700; hemoglobin 85 per cent.

Her mental condition grew steadily worse as shown by incoherence mingled with wild hallucinations and illusions, chiefly of terror, and steady mental reduction. A week or two before her death the optic discs, which were cupped, began to present the appearance of an optic neuritis, but this did not fully develop. About this time her evacuations became involuntary. Later she became stuporous and developed Cheyne-Stokes breathing. She died of all the evidences of cerebral and general physical exhaustion.

Dr. Cadbury, one of the internes at the Pennsylvania Hospital, has kindly furnished notes of the case.

The necropsy was performed by Dr. Geo. S. Crampton, one of the resident physicians to the Pennsylvania Hospital. The results of the gross examination summarized were, chronic interstitial nephritis; chronic mitral and aortic endocarditis; acute vegetative aortic endocarditis; cardiac hypertrophy with fatty degeneration of the myocardium; congestion of the lungs with acute bronchopneumonia; fatty degeneration of the liver, and a high degree of atheroma of the vessels at the base of the brain. The brain was sent to the laboratory of neuropathology of the University of Pennsylvania where it was examined both macroscopically and microscopically by Dr. C. D. Camp under the supervision of Dr. Wm. G. Spiller, professor of neuropathology.

Further macroscopical examination of the brain showed the convexity and other regions of the brain apparently normal. The arteries at the base, as already stated, were intensely sclerotic. The pregeniculæ were much atrophied on both sides. No areas of softening were found in the thalamus, other basal ganglia, optic radiations, or in any part of the cerebrum.

After hardening in ten per cent formalin solution, pieces of tissue from various parts were examined histologically with the following results:

The preparacentral regions on each side were normal by the hematoxylin, acid-fuchsin, and Weigert-hematoxylin methods, though the blood vessels here as elsewhere in the cortex, were slightly thickened and the perivascular spaces enlarged. There was no round cell infiltration of the pia or about the blood-vessels. The Betz cells, studied by the thionin modification of the Nissl method, were intensely pigmented, occasionally almost filled with pigment. The tigroid substance appeared to be normal, except where it was invaded with the pigment. The contour of the cell was preserved, but the dendritic processes of many of the cells were indistinct.

Sections from the cortex of various parts of each hemisphere, selected with a view to their functions, such as the posterior paracentral convolutions, the midprecentral opposite the second frontal convolution, the frontal poles, the quadrate lobules and angular gyres were found to be normal by the acid-hematoxylin, acid-fuchsin, the Nissl, and Weigert-hematoxylin methods.

In sections from the calcarine fissure from each side was seen to be an intense congestion, with the formation of numerous very fine new capillaries. They were mostly in the second and third layers of the cortex, and the pyramidal cells in their vicinity appeared degenerated on examination by the Nissl method. Sections from the cortex of the lateral surface of each occipital lobe, near the occipital pole, showed areas not involving the whole section of a similar vascular appearance, as in the calcarine fissure, and probably a part of the same pathological process. The nerve cells in the location of these areas were of the pyramidal variety, but they had lost their normal shape and structure completely. The white substance beneath these areas appeared degenerated by the Weigert-hematoxylin method, and full of small holes as though nerve fibers had dropped out.

Sections from the white matter in the interior of the frontal lobes appeared perfectly normal by the Weigert-hematoxylin method, but sections of the white matter from the interior of the occipital lobes did not stain so well.

Sections from the oblongata appeared normal by the acid-hematoxylin, acid-fuchsin, and the Weigert-hematoxylin methods. The cells of the nuclei of the hypoglossal nerves appeared normal on examination by the Nissl method of staining.

The left optic nerve was about one-third of the normal size of an optic nerve, and surrounded by a greatly thickened sheath. There was much connective tissue overgrowth between the nerve bundles, which were small and degenerated.

The chiasm appeared to be degenerated by the Weigert-hematoxylin method, but there was no round cell infiltration about it.

It is most interesting to note in connection with this case that the patient was a sister of a lady, the record of whose case was presented to the Medico-Psychological Association in 1897; and also in a fuller report, to the Section on Neurology and Medical Jurisprudence of the American Medical Association in 1898.[2]

In this case the patient was a woman sixty-four years old at the time of her death. At a comparatively early period after the birth of one of her children she had an attack of mania from which she recovered. She had also suffered from chorea during adult life, and when about thirty-five yeare of age began to show some signs of mental change and peculiarity which gradually increased. During the third year before her death she became so unreasonable as to make living with her almost impossible. Later she had attacks resembling grippe. She became bed-ridden and was the subject of persecutory delirium or outbreaks of excitement and of hallucinations of various sort.

Her chief symptoms during the last eighteen months of her life were: vertiginous attacks; difficulty in orientating herself; marked amnesia, not only for names, but for recent events. She gradually became feebler mentally, and during the few months preceding death was in a state of decided dementia, with occasional spells of excitement. Her attempts at conversation were childish, and she had numerous transient, unsystematized delusions.

The necropsy in this case showed on gross examination widespread evidences of arterial disease. The vessels at the base were atheromatous; the precommunicans was the seat of an aneurism, and numerous miliary aneurisms were found in the pial vessels in different locations. The dura was thickened and the pia arachnoid somewhat opaque. An extensive and careful microscopical investigation of six different regions of the cortex was made by Dr. Mary A. Schively, who used the thionin, methylene blue, silver phosphomolybdate, hematoxylin and eosin, hematoxylin, picric acid and fuchsin, and also the Weigert-Pal methods of staining. The results were recorded in a condensed but valuable report with

[2] American Journal of Insanity, vol. 56, No. 2, 1897. The Journal of the American Medical Association, No. 15, vol. xxx, April 9, 1898.

numerous illustrations of the pathological appearances found in nerve cells, glia cells, vessels, meninges, and brain substance. One of the most interesting illustrations was a photo-micrograph of a long pyramidal cell from the ascending frontal convolution, showing roughening of the cell corpus and of the apical dendrites; also moniliform swellings of some of the basal dendrites. The pathological features of the case were summarized as internal and external changes in the neuron; changes in the protoplasmic glia cells; changes involving the cortical pial vessels, also the vessels at the base of the brain; multiple areas of softening in the ascending parietal region; and myelin degeneration.

A close scrutiny of the pathological changes found in the brain recorded in the present article will show that the pathology of the two cases was fundamentally the same. In the first case recorded, however, the disease was much more extensive and more advanced as would be expected from the clinical history of the case, the evidences of mental disorder and deterioration having extended over many years, while in the case of this patient's sister, now put on record, active disease had only been present for a year or two at most. In this case, as in the other, the vessels at the base were highly atheromatous. The cortical vessels were everywhere slightly thickened and the perivascular spaces enlarged. The giant pyramidal cells were in some places almost filled with pigment; and the dendrites, although preserved, were often indistinct. The cortex and subcortex examined in various regions as stated in the full report, showed comparatively little disease. The calcarine cortex, however, and other parts of the occipital lobe were the seat of recent vascular and less recent degenerative changes. The intense congestion and the formation of new capillaries were pathological findings of particular interest in connection with the conditions of excitement and the striking visual hallucinations shown by the patient. The degenerative changes in the pyramidal cells were similar to those found by Dr. Schively in widespread areas in the other case. The pyramidal cells in both cases had lost their normal shape and structure and the adjacent white substance was degenerated. Whether the very marked atrophy of the left optic nerve and the accompanying changes were primary or secondary it is difficult to say. In any case it is probable that the visual hallucinations present were due to the cortical and subcortical vascular alterations.

The case reported in this paper and the one placed on record in the Proceedings of the Association in 1897, in addition to other points of interest, constitute valuable contributions to the symptomatology and pathological anatomy of the insanity of arteriosclerosis. The vessel changes of arterio-sclerotic insanity as recorded by others are, thickened and much altered walls; dilated or occluded lumina; and aneurisms, both miliary and macroscopic. In connection with the intense congestion and formation of new capillaries recorded in our case, it is interesting to note that other cases with similar changes, including capillary hemorrhages, have been recorded. Advanced intracellular, endritic, and neurogliar changes were also present in our case as in others. It is also worthy of note that in the case here first recorded arterio-sclerotic changes were found in other organs as in the kidneys, heart, and liver.

Whether the complete blindness in one eye and the serious loss of vision in the other were to be regarded as constituting a genuine case of crossed amblyopia such as has been reported by Ferrier and Gowers, and attributed to lesion of the angulo-occipital region or of the macular bundle may perhaps be regarded as doubtful. If due to the lesions undoubtedly present in the calcarine cortex, this crossed amblyopia is of much interest in connection with the question of the existence of a separate macular representation in the primary or lower cortical visual centers of the calcarine region and in the higher angulo-occipital visual areas.

Dr. Harlan agreed that the final amblyopia in this case could not be attributed to the preceding glaucoma, at least not to the peripheral conditions which were present. With the exception of the cupped nerves, the fundus of each eye was normal and the filling up of the nerve cup seemed to indicate the occurrence of some intracranial lesion. The attacks of sudden loss of vision which occur in chronic glaucoma may, in some instances be due to intracranial lesions affecting the cerebral, and especially the macular, bundles or centers. It is possible of course that the atrophy of the left optic nerve and tract may have accounted for the great loss of acuity of vision and contraction of the visual field in the left eye, but in any case the sudden amblyopia in the right eye was doubtless due to a sudden vascular lesion in the left hemisphere.

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THE OCCASIONAL CLINICAL RESEMBLANCE BETWEEN CARIES OF THE VERTEBRÆ AND LUMBOTHORACIC SYRINGOMYELIA, AND THE LOCATION WITHIN THE SPINAL CORD OF THE FIBRES FOR THE SENSATIONS OF PAIN AND TEMPERATURE.[1]

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Tuberculous caries of the vertebræ (Pott's disease) is so common that one might think there is little new to say about it, and yet the resemblance it may present to the lumbothoracic form of syringomyelia has not received much attention. There is no doubt that the resemblance between these two diseases in their clinical manifestations at times may be so great that a correct diagnosis may be very difficult. A case of caries of the vertebræ recently under my care presented the symptoms of lumbothoracic syringomyelia in so striking a manner as to make a diagnosis uncertain.

Cases of syringomyelia are numerous in the literature and much has been written about the disease since Schultze and Kahler first recognized the symptoms in 1882 (Mertens). Minor mentions that Schultze, and Kahler and Pick first in 1883 confirmed the clinical diagnosis by necropsy. Usually the cavity is in the cervicothoracic region, although not necessarily confined to this part. It may, however, be limited to the lower thoracic and lumbar regions. Such cases are of rare occurrence. I believe, however, I have seen two or three clinical examples of this type of syringomyelia.

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[1] Read by invitation before the Section on Neurology and Psychiatry of the Medical and Chirurgical Faculty of Maryland, March 8, 1905.

Schlesinger, in his wonderfully thorough work on syringomyelia, remarks that the lumbothoracic type is relatively rare as compared with the cervical type, but its existence has been established by necropsies. Cavities in the lower part of the cord frequently are small and cause no symptoms, but in other cases there may be severe pain and paræsthesia of heat and cold sensations as early symptoms. The spinal column may show kyphoscoliosis early, and may be tender to pressure. Weakness of the lower limbs soon develops, even within a few hours, and may be associated with involuntary jerking movements. Intense atrophy may follow. The lower limbs are usually spastic and the tendon reflexes exaggerated. The symptoms may be confined to one limb a long time, or may develop rapidly in both lower limbs. Girdle sensation is not uncommon. Dissociation of sensation is present, although tactile sensation may be much involved. Ataxia may occur. Vesical and rectal implication are not rare, and may be transitory; the sexual functions also may be affected. Trophic disturbances (mal perforant, cutaneous gangrene, œdema) may be seen in the lower limbs.

As examples of the lumbothoracic type with necropsy, Schlesinger refers to the cases of Lenhossék, Vulpian, Hitzig, Kahler and Pick, and a doubtful case of Oppenheim. He refers to the clinical cases of Lutzenberger and de la Camp. As an example of tumor with cavity within the spinal cord in the lumbar region, the case of Simon (Case VIII.) is mentioned. Clinical cases are referred to as reported by Brunzlow, Oppenheim (2), Mills, Rummo (2), and Tornow.

As examples of clinical cases of the lumbosacral type of syringomyelia (there seem to be none with necropsy) Schlesinger[1] refers to the cases of Oppenheim, Graf, Dercum, Köbner, Gnesda, Minor, Manner, Mertens, and three cases of his own, but in one of his three cases (Case LII.) the upper limbs also were implicated.

It is hard to understand how the diagnosis of syringomyelia could have been made in Lenhossék's case, as given in abstract by Schlesinger, because no dissociation of sensation and no trophic lesions are mentioned. The symptoms were suggestive of meningomyelitis, and the patient had been guilty of excesses, and syphilis might have been suspected. There seems to have been nothing clinically characteristic in Vulpian's case, and the pathological findings, as given by Schlesinger, are like those of meningomyelitis. Hitzig's case could not have been diagnosed during the life of the patient. Kahler and Pick's case or Simon's could hardly have been diagnosed as one of syringomyelia from the symptoms; therefore, there seems to have been no case of syringomyelia in the lower part of the cord with necropsy in which the diagnosis could have been made in life.

Some of the clinical cases mentioned by Schlesinger can hardly be regarded as examples of syringomyelia confined to the lower part of the spinal cord.

In the case reported by de la Camp,[2] trophoneuroses and sensory disturbances of the syringomyelic type in the lower limbs, exaggeration of the patellar reflexes, gradual convexity of the vertebral column without a distinct prominence (gibbus), led to the diagnosis of syringomyelia, but there was no necropsy.

The report of O. Brunzlow's[3] case (Case IV.) is far from convincing, so far as the existence of lumbothoracic syringo myelia is concerned. No necropsy was obtained. The symptoms developed after trauma.

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[1] Die Syringomyelie, Hermann Schlesinger, second edition, 1902 Franz Deuticke, Leipzig und Wien.

[2] Berliner klin. Wochenschrift, March 4, 1901.

[3] Inaugural Dissertation, Berlin, 1890.

The cases reported by McConnell from Dr. Mills'[1] service were interesting, as they were among the earlier cases of syringomyelia reported in America, but they were not of the lumbothoracic type.

E. Tornow's[2] case can hardly be accepted as one of uncomplicated lumbothoracic syringomyelia.

Graf's[3] patient (Case III.) had arthropathies of the left knee and foot, exaggeration of the patellar reflexes, and syringomyelic dissociation of sensation in the lower limbs. The case was regarded as one of syringomyelia, and the diagnosis seems to have been correct, although no necropsy was obtained.

Dercum's[4] case probably was one of syringomyelia of the lower type.

Neither of the two cases reported by Köbner[5] seems to have been an example of lumbosacral syringomyelia, and in each case syphilitic infection had occurred. I should be inclined to regard the pathological process in these cases as syphilitic meningomyelitis.

Max Gnesda's[6] case is surely a doubtful one of lumbosacral syringomyelia.

Minor's[7] case, if I have found the right report, was with necropsy, but was complicated by spina bifida, meningocele, and hydrocephalus.

Simon's[8] case (Case VIII.) was with necropsy and was one of lumbar syringomyelia.

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[1] Journal of Nervous and Mental Disease, 1894.

[2] Inaugural Dissertation, Berlin, 1893.

[3] Beiträge zur klinischen Chirurgie, 1893, vol. x. p. 517.

[4] Journal of Nervous and Mental Disease, 1898, p. 488.

[5] Deutsches Archiv für klinische Medicin, vol. xix. p. 169.

[6] Mitteilungen aus den Grenzgebieten der Medizin und Chirurgie, 1899, vol. iv. p. 744.

[7] Zeitschrift für klinische Medicin, 1898, vol. xxxiv. p. 373.

[8] Archiv für Psychiatrie, 1875, vol. v. p. 156.

Merten's[1] patient probably had had syphilis, and had paræsthesia of the hands and of the occipital region and of the mucous membrane of the mouth; he had also diabetes. The lower limbs were weak and presented the syringomyelic dissociation of sensation. The vertebral column was not affected. The patellar reflex was normal on one side and diminished on the other. Trophic disturbances were present in the feet. Mertens believed the case was one of syringomyelia of the lower part of the spinal cord, but there was no necropsy, and the case was a complicated one.

Thus, without examining all the reported cases, we find that the examples of syringomyelia of the lower type are not numerous. I report the following as a case which should be placed under this heading:

Joanna M., an Italian woman, aged thirty-four years, was admitted to the Philadelphia General Hospital May 11, 1903. The notes I dictated June 3, 1903, are as follows:

A history was obtained, but is unreliable, as the woman replies in the affirmative to almost every question put to her. She is said to have had subjective pain in both lower limbs about four months, but none in the upper limbs. The pain was said to have been first in the right lower limb and developed before the weakness. It is impossible to determine how real this pain was. She had one child seven or eight years ago, and it died two days after birth.

She has less perception of pain in the right lower limb, both thigh and leg, than in the left, as shown by the fact that she does not withdraw the right lower limb from irritation as promptly as she does the left. The right lower limb is not spastic, and, indeed, seems flaccid. The left lower limb is decidedly spastic. There is no patellar reflex on the right side, not even contraction of the quadriceps muscle from tapping the patellar tendon. The Achilles reflex is absent on the right side. The Babinski reflex is present on the right side, but the movement of the big toe is not very marked. The Achilles jerk is exaggerated on the left side. Ankle clonus is not obtained at present examination (it was obtained on the left side five days later).

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[1] Beiträge zur klinischen Chirurgie, 1901, vol. xxx. p. 121.

Babinski's reflex on the left side is very distinct. Both lower limbs are weak, but the left is much weaker than the right. If well supported the woman can take some steps, and in doing so moves the right lower limb forward and drags the left lower limb. She always steps out with the right lower limb.

The upper limbs and face do not appear to be involved. Pressure over the back, she says, is painful in the lower thoracic and lumbar regions, but deep pressure causes no change of expression.

Further notes were made by me December 12, 1903:

The right lower limb is flaccid. The right patellar reflex is completely lost, even on reinforcement. The left patellar reflex is much exaggerated. The left lower limb is spastic, but probably not so much so as at the former examination. Patellar clonus is not obtained on the left side. Ankle clonus on the left side is persistent. No indication of clonus is seen on the right side. Achilles jerk is very feeble on the right side, and it is doubtful whether it is present. Pain sensation is distinctly present in the lower limbs.

February 23, 1905. There are no deformities of the vertebral column, and no trophic disturbances of the lower limbs. Voluntary power in the right lower limb, both in thigh and leg, is about normal. Voluntary power in the left lower limb, both in thigh and leg, is distinctly diminished, but not lost. The lower limbs are not distinctly atrophied.

The patellar reflex is greatly exaggerated on the left side, and there is distinct patellar clonus. Ankle clonus is persistent on the left side. Babinski's reflex is typical on the left side. The patellar reflex is completely lost on the right side, as is also the Achilles jerk on this side. Babinski's reflex is also distinct on the right side, but not quite so pronounced as on the left side. Light touch is perceived promptly in both lower limbs. Pin-prick is perceived in the right lower limb if the prick be deep, but there is undoubtedly diminished pain sensation in the right lower limb. Sensation for heat is much diminished in the whole right lower limb, and probably over the right lower part of the abdomen, although it is uncertain whether there is diminution to heat sensation over the lower part of the abdomen. Sensation for heat is very prompt in the entire left lower limb. Cold sensation is probably diminished in the entire right lower limb. In walking the left lower limb is spastic, and the woman swings it a little from the hip and scrapes the toes of the left foot along the ground very markedly. The right lower limb is thrown unnecessarily far forward in walking, and the heel is brought down first. The left lower limb is a little spastic in passive movements, but the right lower limb is flaccid.

Biceps and triceps tendon reflexes are prompt in the upper limbs, but it is difficult to say whether they are exaggerated. The thenar and hypothenar eminences are well developed in each hand.

The right pupil is a little larger than the left, but the reaction to light is prompt. Sensation for pain seems normal on each side of the face.

A brief summary of this case is as follows:

The condition in this patient has remained unchanged during almost two years. As she cannot understand English and is not very intelligent, I am unable to obtain any account of the development of the disease, even by an interpreter. The symptoms are confined to the lower limbs, if we exclude the inequality of the pupils, and are as follows: In the right lower limb pain and temperature sensations are much diminished; while tactile sensation is prompt, the patellar tendon reflex and Achilles tendon reflex are lost; the limb is flaccid, and its movements in walking seem to be a little ataxic, while voluntary power is about normal. In the left lower limb all forms of sensation are normal, weakness is distinct, the patellar reflex is exaggerated, and patellar clonus and ankle clonus are present; the limb is a little spastic, and its toes are dragged a little when the patient walks. Babinski's reflex is present on each side. It is a Brown-Séquard paralysis (weakness in the left lower limb, dissociation of sensation in the right lower limb), with spasticity and exaggerated tendon reflexes in the left lower limb, and flaccidity and loss of tendon reflexes in the right lower limb.

A lesion in the left half of the spinal cord in the thoracic region and above the lumbar region would explain the weakness of the left lower limb and the exaggeration of the tendon reflexes and the presence of the Babinski reflex in the left lower limb; also the dissociation of sensation (preservation of tactile sensation, with much impairment of temperature and pain sensations) in the right lower limb, because the sensory fibres from the right lower limb cross soon after entering the cord to the left side. We must assume that the crossed pyramidal tract and other parts of the lateral column, especially Gowers' tract on the left side, are affected, but the gray matter of the anterior and posterior horns of this side cannot be diseased in the lumbar region, or there would be atrophy and loss of tendon reflexes in the left lower limb. To explain the loss of tendon reflexes and the flaccidity in the right lower limb without atrophy, we must assume that there is a lesion of the right posterior horn of the lumbar region without involvement of the right anterior horn, as no atrophy has occurred, even after two years, and we should expect atrophy were the anterior horn cells diseased. The lesion may affect slightly the right crossed pyramidal tract, because of the Babinski reflex and possibly slight weakness on the right side, but it cannot greatly impair this tract, as the weakness of the right lower limb is comparatively slight; nor can it involve much of the right lateral column, as sensations of pain and temperature are normal in the left lower limb. The lesion must, therefore, involve much of the left half of the cord in the thoracic region, but not extend into the left lumbar region very far, and must implicate the right posterior horn in the lumbar region; it must also be a slowly developing lesion, and nothing suits these conditions so well as syringomyelia. In the case of syringomyelia reported by Dr. Dercum and myself, the cavity in the cervical region was strictly confined to one posterior horn, although below the cervical region it implicated a large part of the transverse area of the cord.

Cases with symptoms of syringomyelia from fracture I do not consider here, as the history of injury and the signs of fracture make a mistaken diagnosis improbable. Hemorrhage into the cord, occurring as a result of fracture, as shown by Minor, may cause the dissociation of sensation of the syringomyelic type, but this disturbance of sensation may also be caused by myelitis, and in 1903 I[1] reported a case of traumatic myelitis with necropsy without central hemorrhage, in which the dissociation of sensation of the syringomyelic type was present (preservation of, or slight impairment of, tactile sensation, with marked implication of pain and temperature sensations). The lesions in the cervical region in that case were diffuse.

In my experience spontaneous pain or pain on pressure over the vertebræ is not a very common symptom of syringomyelia, although it does occur, and it is mentioned by a number of writers as a sign of the disease. The clinical picture of lumbothoracic syringomyelia we may expect is paralysis or paresis of the lower limbs, with the syringomyelic dissociation of sensation and possibly trophic disturbance in the lower limbs; kyphoscoliosis, especially low in the vertebral column; usually exaggeration of the tendon reflexes of the lower limbs, but sometimes loss of these reflexes, if the cavity involves the reflex arcs, and possibly involvement of bladder and rectum.

It is well to study the literature to see whether the syringomyelic symptom group occurs as a result of lesions other than syringomyelia. The dissociation of sensation of the syringomyelic type I have observed in the case of traumatic myelitis with necropsy referred to above, and also in a case of syphilitic meningomyelitis with necropsy, reported by Dr. C. K. Mills and myself.[1] In these cases the lesions were widespread in a transverse section from the cervical region. It is hard to understand the dissociation of sensation occurring in these cases, but we may conclude that fibres of tactile sensation are more widely distributed in the spinal cord or are more resistant than are the fibres of temperature and pain sensations. These cases do not afford proof for or against the view that fibres of temperature and pain sensations are contained in the tracts of Gowers.

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[1] University of Pennsylvania Medical Bulletin, Feb., 1903.

Minor[2] has shown that a zone of the syringomyelic type of dissociation of sensation may be found just above the level of complete anæsthesia in cases of traumatic myelitis.

Van Gehuchten regards the syringomyelic dissociation of sensation in the lower limbs as exceedingly rare in cases of compression of the spinal cord, and he reports 2 cases of this type, although the diagnosis of the lesion is somewhat doubtful. He refers to a similar case reported by Bruns, to 4 cases reported by Vines, 1 being a case of Pott's disease, and to the case of Pott's disease reported by Edsall, and to the case of compression of the cord reported by Marinesco. In all these cases the syringomyelic disturbance of sensation was in parts of the body innervated below the spinal lesion, while in Minor's cases the dissociation was in the area innervated from the portion of the cord just above the area of compression.

In Marinesco's[3] case of compression of the cord the syringomyelic dissociation of sensation at first was in the lower limbs and in the anterior and posterior parts of the trunk. Later anæsthesia was found in the feet and legs.

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[1] Journal of Nervous and Mental Disease, January, 1903.

[2] Neurologisches Centralblatt, 1898, p. 537.

[3] Semaine médicale, 1898, p. 154.

Dissociation of sensation from pressure of a spinal tumor has been observed (Boettiger,[1] Cushing[2]).

Van Gehuchten[3] believes the dissociation of sensation is dependent upon a lesion of Gowers' tract, and that pain and temperature fibres are contained in this tract. This, he remarked, was a new idea, but if this view could be sustained it would make clear many problems of the pathology of the nervous system. He first expressed in 1893 the opinion that the tract of Gowers contains fibres of pain and temperature sensations. His views have been supported by Brissaud, Lloyd, Schlesinger, and others, but no one has offered any *proof* of the correctness of this opinion. Van Gehuchten has not had any cases with necropsy sustaining his opinion, and in Lloyd's case the lesion was too extensive to afford proof that the tract of Gowers alone contained the fibres of pain and temperature.

One of Van Gehuchten's arguments is that the tract of Gowers is the only long tract in the lateral column likely to transmit fibres of pain and temperature. This is a proper method of reasoning, but sensation possibly may be transmitted by relays of fibres, by connected systems of neurones in the anterolateral columns. Many have held that sensory fibres are probably contained in the anterior or lateral columns; therefore, when the lesion involves much of these columns, we have no proof that Gowers' tract alone conveys fibres of pain and temperature. Petrén,[4] Mai,[5] and Piltz,[6] of still more recent writers, express the opinion that fibres for the sensations of pain and temperature are contained within Gowers' tract.

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[1] Archiv für Psychiatrie, vol. xxxv, p. 83.

[2] Annals of Surgery, June, 1904.

[3] Semaine médicale, 1899, p. 113, and Journal de neurologie, July 20, 1897, p. 262.

[4] Cited by Mai.

[5] Archiv für Psychiatrie, 1901, vol. xxxviii. p. 182.

[6] Neurologisches Centralblatt, March 16, 1905.

The case of caries of the vertebræ presenting symptoms of lumbothoracic syringomyelia I have had the opportunity to study is as follows:

John R., colored, aged twenty-three years, was admitted to my service in the Philadelphia General Hospital August 31, 1904, and, after passing into Dr. Mills' service, came again under my care. He died January 13, 1905. Chief complaint August 31, 1904, pains in legs, abdomen, and back. Last January he fell from a ladder and injured his left hip slightly. He gives a history of gonorrhœa last summer, but denies specific disease.

He began four months ago to have dull pains in the lumbar region of the back and abdomen and on the left side. Then numbness in the feet developed and gradually extended up both legs. He has gotten weaker generally during the past two months. When he elevates his arms it causes pain in his back. He has not felt girdle sensation. At times he gets dizzy and staggers. Sight and hearing have not been affected. He is a fairly well-developed male. He has a number of scars on the abdomen, which he received in childhood as a result of of burns, which he says were not painful. The lungs appear to be normal.

I made the following notes September 1, 1904:

There is marked deformity of each knee which is not very great while he is sitting, but when he is standing he has extreme genu valgum, such as would occur from a relaxation of the ligaments. He has no pains in his limbs, but the lower limbs from the knees down feel as if asleep. The deformity of the lower limbs has existed only about five years. One brother and three sisters of the patient are living and have no deformity of the limbs. The man is not emaciated. The patellar reflexes are a little exaggerated on both sides. Resistance to passive movements in the lower limbs is slightly diminished. Sensation for touch in the lower limbs is normal or nearly normal. Sensation for pain is abolished or nearly abolished in the lower limbs. Sensations for heat and cold are almost lost in the lower limbs below the knees, much impaired in the thighs and lower part of the trunk, and are normal in the upper limbs.

Sometimes he cannot hold his urine, but has no difficulty in passing it. His bowels are not usually constipated. When standing his feet are far apart, because of the deformity of his knees. He sways very slightly with his eyes open, but sways distinctly when they are closed. The gait is difficult because of the bilateral genu valgum. The Achilles jerk is absent on each side. The Babinski reflex is not present on the left side, the movement of the toes not being positive in any direction. It seemed to be present on the right side two or three times, and then was exhausted. He is flat-footed on each side. No girdle sensation is felt. There is a very slight kyphosis in the lower thoracic region about the tenth thoracic vertebra with slight scoliosis.

The upper limbs are normal.

The pupils are equal; the irides contract promptly on both sides. The cranial nerves are not involved. Sensations in the face for touch, pain, and temperature are normal. Seventh, twelfth, and fifth nerves are normal, as are the extraocular muscles.

On September 16, 1904, he fell down-stairs backward, was unable to pick himself up, and was carried to bed, where he has remained. Immediately after the accident he was unable to stand or walk, and had considerable pain in the lumbar region.

Dr. McConnell made the following notes: October 2, 1904:

He is almost completely paraplegic; slight evidence of attempt to move the limbs is observed in contraction of the quadriceps. When the limbs are supported under the knees the patient can kick slightly. He is unable to dorsally flex either foot. Sensation to touch seems to be lost in the whole of the right leg, but is quite well preserved in the left leg, penis, and scrotum, and everywhere else in the body. Sensation to temperature is lost everywhere over the right leg and foot; also over the left foot, but preserved everywhere else. (It is not said that pain and temperature sensations were normal in other parts.) Sensation to pain seems to be lost and preserved in the same distributions as are the other sensations. The patellar reflex is very markedly increased on each side. Ankle clonus is obtained on each side. Babinski's sign is not positively obtained, although there seems to be an occasional abnormal response to plantar irritation on each side. Achilles jerk is lost on each side. Gastrocnemius and quadriceps jerks are both retained. Squeezing the testicles does not cause any pain. Cremaster reflex is questionable.

October 17th. He complains of pain on both sides of the abdomen and in the back. Percussion at the base of either side of thorax causes pain. There is a dull area on the right side, extending from the fifth rib to the costal margin. Left side is resonant.

27th. Sensation to touch seems to be everywhere present in the right limb, as well as in the left; even the soles of the feet seem to respond to touch stimulation. Pain sense is irregularly present, the response to pain stimulation being very uncertain and not always correct, although for the most part he *seems* to have pain sensation. Temperature stimulation, while better responded to than formerly, is not always correctly responded to. He will speak of hot as cold and cold as hot within very small areas on the surface. He is unable to tell which of the toes is grasped on the right side, but at once responds when the left foot is examined in a similar manner.

He has developed three bedsores within the last few days.

November 30th. Temperature runs up at night, and he sweats rather profusely. He cannot move his legs at all and complains of pain in the right knee. Patellar and ankle clonus are obtained on both sides. Exaggerated knee-jerks and Babinski's reflex are present on both sides. Tactile sensation is dulled in the legs, and he has some paræsthesia.

December 10*th*. The man says he can hold his urine well, and has to percuss the abdomen in order to start the flow of urine. Bowels are constipated. He is completely paraplegic; cannot move even a toe. Knee-jerks are both markedly increased. Patellar clonus is marked on each side. Ankle clonus and Babinski's reflex are present on each side. Tactile sensation is preserved on the right side. He seems unable to appreciate the difference between heat and cold over both legs generally. He almost always responds correctly to the application of cold, but is unable to tell when heat is applied until a point is reached on the inner surface of both thighs corresponding to the riding breeches' area. The scrotum and penis are sensitive to heat and cold. On the posterior surface of both thighs the response to heat again corresponds in its lower border to the riding breeches' area. Above this lowest line sensation seems to be normal. The trunk and upper extremities seem to be normal in response to all forms of sensory stimulation. The legs are considerably wasted, particularly in the inner side of the thighs and the calf muscles of the right leg; here, however, the wasting is relative as compared with the left leg, both calves being quite small. No wasting is seen in the feet, but there is a very marked degree of desquamation, probably trophic, more on the left leg, especially the outer surface.

In January, 1905, the patient came again under my care.

January 8, 1905. He has been having increased temperature and is becoming more emaciated, but has no cough. There is great wasting of the thigh and calf muscles. He complains of pain in the legs, but no girdle sensation. Incontinence of feces is present, although he is generally constipated, but has good control over the bladder. Sensation over the legs is felt for strong pin-prick, but is impaired or lost for light touch; sensation in the right leg being sometimes referred to the left.

11*th*. He is much weaker and almost stuporous. Abdomen is tympanitic, and there is a nodular mass palpable in the left umbilical region, and the same less evident to the right slightly higher up. Twitching of arms and fibrillary tremors of face are observed. Incontinence of urine and feces have been present since yesterday.

12*th*. He is comatose and lies with his head inclined to the right and persistently refuses to turn it to the left. He has irregular feeble movements of both upper limbs and fibrillary twitching of facial muscles of both sides, especially of the right side. Head is held very stiffly to the right. Eyes are held also far to the right. The right upper limb is moved freely when stuck with a pin; the left upper limb is moved only slightly when it is stuck, and is paretic.

When the face is stuck with a pin contraction of facial muscles follows on each side. Pin-prick is certainly felt, both in the face and upper limbs. Passive movement of head to left causes much pain. Sticking either lower limb with a pin causes no movement. Plantar reflex is very much exaggerated on each side. Babinski's reflex is present, but somewhat indistinct on the right side, and uncertain on the left. The patellar reflex is present, but very weak on each side, not causing any movement of the leg, and is detected only by contraction of the quadriceps. It is impossible to determine whether the left leg is more paralyzed than the right, because when either leg is allowed to fall it drops lifeless. Some tendency to flexion contraction of each upper limb is seen at the elbow, especially on the left side.

The necropsy was made January 14, 1905, by Dr. John Funke, whose diagnosis is as follows:

Miliary tuberculosis of lungs, spleen, kidneys, liver, and of right pleura; fibroid myocarditis, spinal caries, large psoas abscess, external spinal pachymeningitis.

My microscopic study of the spinal cord shows a tubercle in the right lateral column at the extreme lower end of the thoracic cord and involving the area of Gowers' tract. About one-half to one inch higher another tubercle is found in the left Gowers' tract. These tubercles are small in all diameters.

The cord does not show any compression from the external pachymeningitis.

Sections from the lumbar region show marked degeneration of each crossed pyramidal tract. The inflammation of the pia here, as everywhere, is very intense. There is also a round-cell infiltration about the vessels within the cord and within the nerve roots external to the cord. Marchi sections show distinct recent degeneration almost confined to the crossed pyramidal tracts. The nerve cells of the anterior horns stained by thionin appear to be normal.

Sections from the lower thoracic region show intense recent degeneration in the crossed pyramidal tracts, and also diffuse recent degeneration in other parts of the transverse section. Here and there in all the white columns are holes where nerve fibres have disappeared. Here also perivascular cellular infiltration within the cord is pronounced. The tubercles are degenerated at the centre and contain giant cells.

Sections from the upper thoracic and cervical regions stained by the Weigert hæmatoxylin method or acid fuchsin do not show distinctly secondary degeneration, but here, as everywhere in the cord, the pia is greatly invaded by round cells, and there is also some perivascular cellular infiltration within the cord.

Marchi sections from the cervical swelling show recent degeneration in the columns of Goll, the direct cerebellar tracts, and the tracts of Gowers. The remainder of the anterolateral columns seems to be almost entirely free from recent or old degeneration. The nerve cells of the anterior horns in the cervical region by the thionin stain seem to be normal

The cellular infiltration of the pia over the medulla oblongata and about one of the optic nerves is as intense as over the spinal cord, and there is slight perivascular cellular infiltration within the medulla oblongata.

A brief summary of this case is as follows: A man, aged twenty-three years, began to have pain in the lumbar region and abdomen four months before he came under my observation. Numbness was soon felt in the feet, and gradually involved the lower limbs below the knees. When standing he had extreme genu valgum, which had existed about five years. He had scars on the abdomen, which he said were the result of painless burns in childhood. Resistance to passive movements was slightly diminished in the lower limbs. Sensation for touch was normal or nearly normal in the lower limbs, but sensation for pain was abolished or almost abolished in these parts. Temperature sensation was almost lost in the lower limbs below the knees, and much impaired in the thighs and lower part of the trunk. Walking was difficult because of the deformity of the knees. He had a slight kyphosis at about the tenth thoracic vertebra. The upper limbs and face were not affected.

After a fall backward down a flight of steps he became completely paralyzed in the lower limbs. Sensation for touch was then lost in the right lower limb for a time, but was preserved in the left lower limb. Sensations for temperature and pain were lost in the right leg and foot and in the left foot, but were preserved elsewhere, although it is not stated that they were normal elsewhere. The patellar reflexes were much exaggerated, and Babinski's sign was present.

Still later sensation for touch was found preserved everywhere in the lower limbs. Sensation for pain was irregularly present in the lower limbs, the response to pain stimulation being very uncertain, and sensation for temperature was also much impaired in the lower limbs.

Tuberculous meningitis and caries of the vertebræ were found, and a small tubercle was present in the right lateral column at the extreme lower end of the thoracic cord, involving the area of Gowers' tract. About one-half to one inch higher another small tubercle was found involving the left tract of Gowers. Although there was some myelitis, it was very evident from the symptoms that it had chiefly developed after the fall down the flight of steps, as at the time the patient first came under my observation weakness in the lower limbs was slight. He lived four months after the accident.

The resemblance to lumbothoracic syringomyelia in this case consists in the scars on the abdomen received in childhood as a result of painless burns; the preservation of tactile sensation, with almost complete loss of pain and temperature sensations in the lower limbs, although the degree of this dissociation of sensation varied when the patient was at forced rest in bed; the extreme deformities of the knees, genu valgum, of five years' duration and resembling closely arthropathies; the slight exaggeration of the patellar reflexes, and the slight diminution of voluntary power in the lower limbs before the fall; the very slight kyphoscoliosis found only by careful examination; the absence of signs of psoas abscess, and of tuberculosis of the lungs before the accident, and the

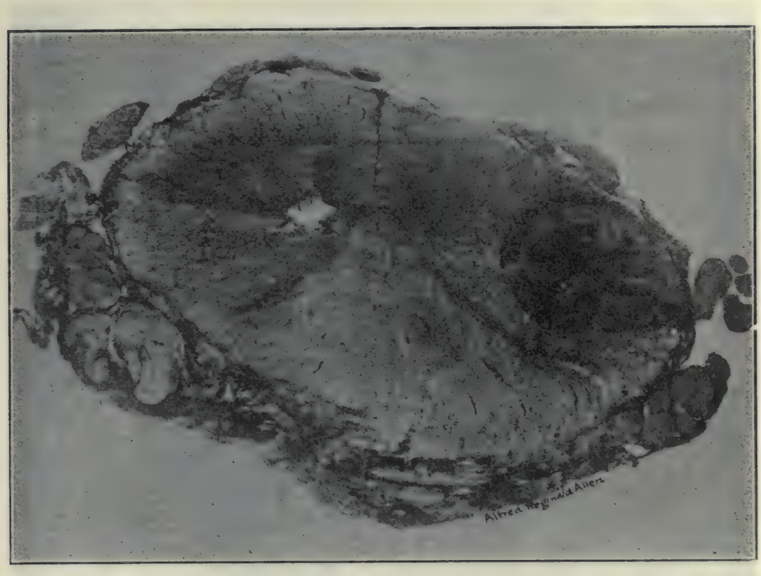

FIG. 1.—Small tubercle in the right lateral column, extending forward into Gowers' tract.

absence of a pronounced kyphosis. This, however, is not the first case of caries of the vertebræ without distinct kyphosis I have seen.

The attack of unconsciousness, with conjugate deviation of the head and eyes to the right, and the paresis of the left upper limb, developing a day or two before death, were the result of intense tuberculous cerebral meningitis.

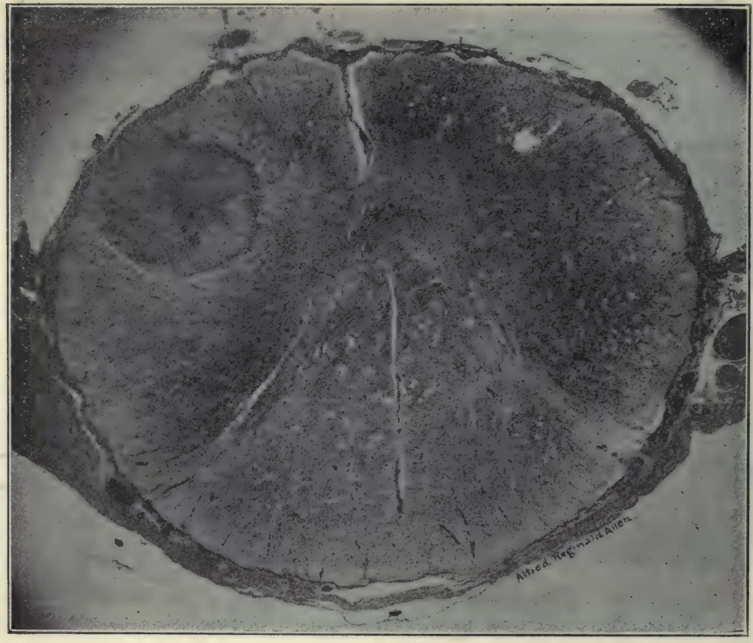

FIG. 2.—Small tubercle in the left anterolateral column, implicating Gowers' tract.

The attempt has usually been made to explain the dissociation of sensation occurring in syringomyelia as a result of a cavity in the gray matter, or when this symptom occurs from other causes the lesion, whatever it may be, has been placed in the gray matter. This seems proper, because, so far as possible, we try to explain all the symptoms in a case by one lesion, although occasionally we err in so doing. A lesion in each anterolateral tract, if it involves the area in which the fibres of temperature and pain sensations are contained, must cause dissociation of sensation, just as truly as a lesion cutting sensory fibres crossing in the centre of the spinal cord, and the former is what has occurred in my case of caries of the vertebræ reported in this paper. The tract of Gowers is invaded by a tubercle on each side, which extends not much beyond this tract on each side, and this case seems to afford proof that Gowers' tract contains fibres of temperature and pain. We can understand why temperature and pain sensations are so often involved in the Brown-Séquard type of paralysis and tactile sensation escapes. The most common cause of spinal hemiplegia, which is of the Brown-Séquard type, is spinal syphilis, and the extension of the syphilitic inflammation from the pia into the peripheral areas of the cord and involvement thereby of Gowers' columns is of frequent occurrence. I believe that tactile fibres, at least in part, are in the posterior columns.

It seems very strange that after my patient had received a severe fall, had thereby become completely paralyzed in the lower limbs, and had been confined to bed, sensations for temperature and pain in the lower limbs should have improved during a brief period, although later they again became seriously affected. It may be that a few axis cylinders passed through the tubercles in the tracts of Gowers undamaged and became capable of transmitting some impressions for a time when the patient was at rest. The existence of normal axis cylinders within a tubercle has been shown. The spinal cord presents now diffuse degeneration, but this probably occurred after the severe fall the patient had, when he became entirely paralyzed. At the first examination I made, the lower limbs were not very weak, and the dissociation of sensation was typically syringomyelic in type, and was evidently caused by the two tubercles, one in each tract of Gowers, because these lesions are of long duration, and degeneration has occurred in the centre of each tubercle.

The severe fall, without causing fracture of the vertebræ, started a partially latent process and caused thereby complete paralysis of the lower limbs. The diffuse degeneration may, with propriety, be considered as of recent development, for it is especially intense in the crossed pyramidal tracts, but at my first examination very little weakness of the lower limbs was detected.

In a paper published in 1898 I[1] mentioned that Strümpell[2] takes a very decided stand in regard to the presence of meningomyelitis in Pott's disease. He says that all the changes in the spinal cord in spinal caries are only the necessary consequences of purely mechanical pressure. "At all events," he continues, "as we must state after numerous examinations made by us, in opposition to the prevailing view, no one has the slightest right to explain the paralysis occurring from spondylitis as the result of secondary myelitis."

If preparations are made from the spinal cord, there are, according to Strümpell, no vascular changes to be found, no hyperæmia, no accumulations of cells about the vessels, and only occasionally small traumatic hemorrhages. Frequently there are foci of degenerating nerve fibres, groups of swollen axis cylinders, with partial or complete loss of their medullary sheaths, and in places spaces from which the nerve fibres have disappeared. In later stages there is proliferation of the neuroglia.

Kraske[3] has expressed similar views in saying that the paralysis cannot be attributed to secondary inflammatory changes in the cord, and that this is equally true of the rare cases in which the cord is involved in the tuberculous inflammation, for this involvement occurs after the paralysis and toward the end of life.

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[1] Johns Hopkins Hospital Bulletin, June, 1898.

[2] Lehrbuch der spec. Path. u. Ther., vol. iii. p. 197.

[3] Archiv für klin. Chirurgie, vol. xli.

I share the opinion that tuberculous myelitis is not a common complication of caries of the vertebræ, but the case I reported in 1898 and the case that is described in my present paper are proof that occasionally it may occur.

PRESIDENTIAL ADDRESS.

THE IMPORTANCE IN CLINICAL DIAGNOSIS OF PARALYSIS OF ASSOCIATED MOVEMENTS OF THE EYEBALLS (BLICKLAEHMUNG), ESPECIALLY OF UPWARD AND DOWNWARD ASSOCIATED MOVEMENTS.

By William G. Spiller, M.D.,[1]

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To preside over this distinguished body of American neurologists is an honor that your chairman feels too deeply to express adequately in words.

"* * * like a cipher,
Yet standing in rich place, I multiply
With one 'We thank you' many thousands more,
That go before it."

Our membership list now is almost full, and we shall be obliged to raise the number above one hundred or to reject desirable applicants. There is some danger in doing the former. When there are many vacancies a candidate is more likely to be accepted without careful scrutiny of his record than when admission is of necessity limited. On the other hand it would be unfortunate to bar out desirable men. A moderate increase in our membership may be advantageous, and the Council will suggest the placing of the limit at 110.

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[1] Read at the meeting of the American Neurological Association, June 1, 1905. From the Neuropathological Laboratory of the University of Pennsylvania.

It is proposed to make some important changes in the Congress of American Physicians and Surgeons. The subject has already come before this Association but must be considered again. The following resolution is offered: Resolved: That in the opinion of this Society it is desirable that the Executive Committee of the Congress of American Physicians and Surgeons of 1907 should consider the advisability of altering the constitution of the Congress, and report progress thereon to the several Societies interested at their sessions of 1905 and 1906.

In preparing this address it has seemed best to me to follow a custom that on occasions such as this has been more common abroad than at home, and to discuss a subject of general interest. I therefore present for your consideration the results of some investigations that may in a small degree increase our knowledge of the disorders of the nervous system.

Disturbances in the associated movements of the eyes (Blicklähmung) afford a sign of localizing value in lesions of the brain, and yet this subject has received comparatively little attention. V. Kornilow, who has written the best recent paper on these palsies, remarks that clinical cases carefully studied are rare, and even more so are those with necropsy.

The opinion of Parinaud and Sauvineau that upward or downward associated ocular paralysis is always associated with paralysis of convergence is incorrect, as shown by a case reported by Parinaud himself, by a case reported by v. Kornilow, and by other cases. It is true, however, as Parinaud pointed out in 1883, that in paralysis of associated ocular movements diplopia often is absent. Indeed when in such palsy diplopia later appears it may be a sign of improvement, as it was in one of the cases of my series (Case 5), where the disappearance of the palsy in one eye caused diplopia, soon followed by complete recovery. Although paralysis of associated ocular movement of any form is usually a sign of grave significance, one should be careful about giving a fatal prognosis. In a case reported by Raymond and Cestan[2] the paralysis lasted ten years, and in a number of cases complete recovery has occurred (Case 5 of my series, and other cases), while in others improvement has been very marked (Case 8 and Case 9, of my series).

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[2] Raymond and Cestan. Revue Neurologique, 1901, p. 70.

Having had the opportunity recently, through the kindness of many colleagues, to whom credit is given in the detailed reports, to study four cases (Beier,[3] Cases 1, 2 and 3), in which paralysis of lateral associated ocular movements occurred, and nine cases in which paralysis of upward or downward associated ocular movements was a striking sign (including three of the four cases mentioned above); having also examined microscopically the material obtained from four of these cases, I have selected paralysis of associated ocular movements as the subject of my address. I shall not devote much space to the study of the paralyses of lateral associated movements, as these have received much attention, and I[3] have recently discussed this subject. The evidence is strong that this form of paralysis is indicative of a lesion of the posterior longitudinal bundle near the sixth nucleus. The external rectus muscle on the side of the lesion may be more affected than the internal rectus of the other eye, and this according to Parinaud[4] is the rule. He mentions also that the movement of convergence may be greater in the eye in which the external rectus is paralyzed, because of the paralysis of the external rectus.

The disturbances of the upward or downward associated movements have been studied less than those of lateral associated movement. The former, though usually acquired, may be congenital, and Oppenheim[5] has observed congenital paralysis of associated upward movement. Uhthoff[6] speaks of paresis of upward associated movement with slight impairment of movement in other directions, occurring in multiple sclerosis.

We have reason to believe that the cerebral cortex exerts some control over the associated movements of the eyeballs; especially strong is the evidence concerning lateral conjugate movements. Conjugate deviation of the eyes has been studied within the past few years by Klaas,[7] and from his investigations of the literature, it appears that this phenomenon was first observed by Andral (1834). It was studied by Duplay, Durand-Fardel and others, but Prevost showed that conjugate deviation of the eyes often occurs in hemiplegia and has nothing to do with strabismus, as Andral believed; he showed further that it is often associated with deviation of the head; that the deviation is toward the lesion when the lesion is cerebral, and away from the lesion when it is in the isthmus. Landouzy believed that irritation and paralysis produce deviation in opposite directions when the lesion is in the cerebrum, and Grasset showed that the patient looks at his paralyzed limb when the lesion is a paralyzing one in the brain stem, and at the lesion when it is an irritating one, and in this part of the brain. Wernicke assumed that the rapid disappearance of conjugate deviation of the head and eyes is owing to the partial control of the lateral ocular movements by the cerebral hemisphere of the same side, and to the action of this hemisphere in place of the injured opposite cerebral hemisphere. Wernicke believed that conjugate deviation of the eyes is caused by a lesion of the lower parietal lobe, and he reported three cases of lesion in the lower part of each parietal lobe in which voluntary lateral associated ocular movements were lost, pseudo-ophthalmoplegia, although in another case with similar lesions disturbance of the ocular movements was not observed.

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[3] Potts and Spiller. Univ. of Penna. Med. Bul., Dec., 1903.

[4] Parinaud. Archives de Neurologie, Vol. 5, No. 14, 1883, p. 145.

[5] Oppenheim. Lehrbuch der Nervenkrankheiten, 4th Ed., Vol. 2, p. 706

[6] Uhthoff. Archiv. für Psychiatrie, Vol. 21, 1890, p. 379.

[7] W. Klaas. "Ueber konjugierte Augenablenkung bei Gehirnkrankheiten." Inaugural Dissertation, Marburg, 1898.

The evidence concerning a center for lateral movements of the eyes in the lower parietal region or angular gyrus is conflicting. There is more evidence that a center for lateral associated ocular movements is situated in the posterior part of the frontal lobe.

The recent investigations of Grünbaum and Sherrington by electrical irritation have again demonstrated that a center for ocular movements probably exists in the frontal lobe. Pathological alteration of the frontal lobe has given evidence to the same effect. In a case of cerebral syphilis with severe lesion of the right frontal lobe, reported by Karl Schaffer,[2] paralysis of associated movement of the eyeballs to the left occurred, and Schaffer concludes from a study of this case that a center for lateral associated movements exists in the posterior part of the second frontal convolution, although he does not deny that there may be another and similar center in the supramarginal and angular gyri.

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[2] Schaffer. Neurogisches Centralblatt, Nov 16, 1904. p. 1035.

Hemianopsia cannot be regarded as the cause of conjugate deviation in all cases, although much has been attributed to it recently by certain French writers. Dejerine and Roussy[9] have demonstrated that it is not necessarily the cause of the deviation, inasmuch as deviation existed in a patient of theirs who had been born blind and whose cortical visual zone had not been developed.

So far the knowledge gained relates chiefly to associated lateral movements, but we know much less concerning a cortical center for upward or downward associated movements.

The views of Parinaud and some other writers on this subject lack confirmation. They believed that there is a center for ocular movements in the occipital lobe and a more important one in the frontal lobe. The inferior part of the latter center controls the upward movement of the eyes, the superior part controls the downward movement of the eyes, the intervening part controls the lateral movements. It is true that lateral associated movements have been impaired with paralysis of upward or downward associated movements in a number of instances, as they were in some of my cases, but a lesion in the vicinity of the corpora quadrigemina will better explain this form of paralysis than a lesion of the cerebral cortex. Teillais says regarding his own case, it would be unreasonable to regard the lesion as cortical, because if the lower part of the cortical center controls the upward movements, and the upper part the downward movements, then the whole of this center except the middle portion must have been destroyed, as the lateral movements were preserved. Such a view cannot be accepted.

Paralysis of lateral associated movement from cortical lesions, as seen in the conjugate deviation of the eyes, is always, so far as I know, transitory, and the persistence of such paralysis points to a lesion of the posterior longitudinal bundle. We have little or no anatomical or pathological evidence that is really valuable concerning the existence of a cortical center for upward or downward associated movements, but in reasoning from analogy we must assume that such a center or centers exist. The clinical case that Parinaud[10] reported in 1892 as indicating a lesion of the frontal center, is in the light of our present knowledge unsatisfactory because of the association of homonymous lateral hemianopsia.

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[9] Dejerine and Roussy Revue Neurologique, Feb. 15, 1905, p. 161.

Parinaud reported the case before the Society of Ophthalmology, April 5, 1892. The man had paralysis of the lower left part of the face, of the left half of the tongue, and temporarily of the left hand. He had almost complete paralysis of the elevators of the eyeballs, with a little reduction in the extent of the lateral movements. Convergence was well performed. (Here Parinaud presents a case of paralysis of upward associated movement with preservation of convergence.) Downward movement was normal. Parinaud believed there was a lesion in the right frontal lobe, at the level of the lower part of the Rolandic fissure, involving the centers for the face, tongue, hand, and associated movements of the eyeballs.

This patient had transitory partial left homonymous hemianopsia, and this was supposed to be the result of a lesion near the visual zone, which occupies, according to the teaching of Parinaud in 1892, all the occipital lobe, and extends to near the ascending parietal convolution. This opinion we now can hardly accept.

Hysteria may cause paralysis of the associated ocular movements, but there is no pathognomonic sign of this form of the palsy, and in some instances a correct diagnosis may be extremely difficult or even impossible. No case presents this difficulty more clearly than the one reported by Crouzon, Marie and Babinski[11] before the Neurological Society of Paris on three different occasions in 1900 and 1901.

Crouzon presented the case as one of spasmodic tic, later Babinski and Parinaud expressed the opinion that it was organic and caused by a supranuclear lesion, and still later Marie presented the case with the diagnosis of a purely functional disorder, a neurosis, as a spasm of the elevators of the eyeballs. When the patient threw his head as far backward as possible and followed with his eyes a finger lowered slowly the eyeballs moved downward, but if his head were erect and he attempted on command to look at his feet, he bent his head forward and the eyeballs went forcibly upward. After thirty or forty seconds the spasm disappeared and the eyeballs returned to the normal position. Marie from this concluded that there must be a spasm of the elevators and not a paralysis of the depressors. The disorder had occurred after an apoplectic attack lasting seventeen hours, and the man had become blind. During the attack he had not had stertorous breathing nor passage of urine or feces, and this Marie regarded as remarkable if there were an organic lesion. The coma disappeared suddenly, but the patient was delirious for several weeks and did not recognize anyone or his surroundings. This Marie regarded as like hysteria. The speech was slow and hesitating, and the visual fields were contracted.

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[10] Parinaud. Annales d'Oculistique, Vol. 107, p. 283, 1892.

[11] Revue Neurologique, 1901, p. 428.

Parinaud pointed out that the downward movement in following a finger is a reflex act and better performed than a voluntary movement, and this he says he has observed in hysteria, but when the condition is hysterical there is no inconvenience from the ocular disturbance, just as there is no inconvenience from the contraction of the visual fields in hysteria, and Parinaud has never seen a hysterical patient incline the head forward in order to make use of the superior part of the *champ du regard* (Dr. de Schweinitz suggests as a suitable translation for this term, field of fixation). The dissociation between the voluntary and reflex movements is not always hysterical. Parinaud has pointed out that spasm may occur in sound ocular muscles as the result of paralysis of other ocular muscles when the patient is required to look in different directions. A paralysis confined to one eye may be the cause of spasm in the other, so that the sound muscles may appear to be affected. In these cases the paralysis is peripheral, but even in associated palsies it is not rare to see disturbance of the antagonistic muscles. In conjugate lateral paralysis from lesion of one sixth nucleus there is almost always nystagmus when the patient looks toward the sound side, and this nystagmus may be so pronounced that it may be difficult to say which side is affected. This I also have noticed in a case of complete paralysis of lateral associated movements on one side and of partial paralysis on the other. In the case which called forth so much discussion in the Neurological Society of Paris the visual fields were contracted only in the inferior portion and not concentrically, therefore the contraction was not like that of hysteria. The *champ visual* (visual field) Parinaud distinguishes from the *champ du regard* (field of fixation). He believes that if the cortical center for associated movements of the eyes in the frontal lobe be destroyed, the voluntary associated movements will be lost, but the reflex associated movements will be preserved, and yet little, if any, clinical evidence in support of this opinion can be found.

Babinski pointed out that in the case under discussion the downward movement of the eyes was imperfect with the head thrown back. He quoted the case reported by Schröder[12] in which only paralysis of the associated downward movement persisted, and the eyes could not be lowered below the horizontal plane. Any attempt to look downward caused a spasm of the elevators and eyes turned upward.

Oppenheim[13] has observed cases of pseudobulbar paralysis in which lateral movements of the eyeballs were impaired in voluntary innervation, but were preserved when the patient tried to follow an object or to turn toward the direction from which a sound came.

All the pathological evidence that I have been able to obtain in cases of persistent palsy of associated upward or downward movement is indicative of a lesion near the aqueduct of Sylvius. It is extremely doubtful whether a lesion confined to the corpora quadrigemina and causing no pressure on the surrounding parts ever causes paralysis of associated ocular movements, and those who favor such a view have not produced proof of a supranuclear center in this part. Bernheimer[14] says Topolanski showed that electrical irritation of the corpora quadrigemina of rabbits does not produce ocular movements, and that these parts could be removed without producing symptoms. He believed that there is a center for ocular movements near the oculomotor nucleus.

Bernheimer, from experiments on monkeys, has shown that the ocular movements do not depend on the integrity of the corpora quadrigemina, that this part of the brain does not contain a reflex center for these movements, and that after destruction of this part the ocular movements are normal. He obtained synergic ocular movements from irritation of the angular gyrus both before and after he had removed the corpora quadrigemina. He thus refutes Prus' statements that a center for ocular movements exists in the corpora quadrigemina.

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[12] Schröder, quoted by Teillais, Bul. de la Soc. Fran. d'Ophthal., 1899, p. 415. (See my abstract of this case.)

[13] Oppenheim. Neurologisches Centralblatt, 1895, p. 40.

[14] Bernheimer. Wiener klin. Wochenschrift, No. 52, 1899, p. 1310.

Cases have been reported in which the corpora quadrigemina have been destroyed without disturbance in the movements of the eyeballs (Weinland, Seidel, Ruel, Nissen, cited by v. Kornilow).

In Case 2 of my series there was complete paralysis of right associated lateral movements, and great impairment of left and of upward associated movements. The paralysis of lateral associated movements is to be explained by the involvement of both posterior longitudinal bundles. If we assume that the nuclei of the superior rectus and inferior oblique muscles are in the posterior part of the oculo-motor nucleus, we can understand why marked impairment of upward movement was present in this case, but this is contrary to the views held by some concerning the position of these groups of nerve cells.

Siemerling[15] has recently remarked that the numerous experimental studies of Bach, Bernheimer, van Gehuchten and van Biervliet have not led to uniform conclusions. The oculomotor nucleus is merely the place for the transference of impulses and we are not in a position to state which parts are concerned with the innervation of the individual muscles.

My Case 1 is further evidence that the nuclei of the superior rectus and inferior oblique muscles may be in the posterior part of the oculomotor nucleus, because there was paresis of upward associated movement and the nerve cells of the oculomotor nuclei were not diseased, but the posterior part of the nuclei was affected by pressure from the tumor in the pons, and the aqueduct of Sylvius was compressed upon the left side. The tumor did not invade the nuclei of the third and fourth nerves. The paralysis of left associated lateral movement is explained by the almost complete destruction of the left half of the pons by a tubercle, and thereby involvement of the posterior longitudinal bundle. The left sixth nerve was also much degenerated. In both Cases 1 and 2 downward movement was normal, this would seem to be evidence that the nucleus of the inferior rectus muscle must be in advance of those for the superior rectus and inferior oblique muscles, as in each case the tumor grew from the pons and did not invade the oculomotor nucleus.

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[15] Siemerling. Archiv. für Psychiatrie, Vol. 40, No. 1, p. 61.

If this is the correct position of these nuclei, a lesion developing ventrally in the pons as shown in two of my cases may by pressure upon the posterior part of the oculomotor nuclei cause paralysis of associated upward movement, as a result of injury of the nuclei for the superior rectus and inferior oblique muscles, and it would not be at all necessary to assume the existence of a coordinating center for upward associated movement. Paralysis of downward associated movement depends on impairment of the inferior rectus and superior oblique muscles, these having cells of origin in two distinct nuclei. There must be a close association between the nucleus of the superior oblique and that of the inferior rectus muscle of the same side, and possibly of the opposite side. It is probable therefore that a lesion of the nuclei of the inferior rectus muscles and of the fibers connecting them with the nuclei of the trochlear nerves would cause paralysis of associated downward movement, and it seems unnecessary to assume that a coordinating supranuclear center for upward or downward movements exist in the corpora quadrigemnia or their vicinity. In deed two of my cases (Cases 1 and 2) show that such a center is improbable, because in each case the lesion causing paresis of upward associated movement was posterior to the oculomotor nucleus, and did not implicate the corpora quadrigemina. Three of the cases of my series afford further evidence against a center for coordinate ocular movements in the corpora quadrigemina. In one the paralysis at first (Case 4) was only in upward movements, later also in lateral associated and in downward associated ocular movements. In this case therefore a paralysis at first confined to one form of associated movement later extended to other forms. In another case (Case 5) the paralysis of upward associated ocular movements disappeared, leaving only paresis of the left superior rectus, and finally this also disappeared. In Case 9 the associated palsy disappeared, leaving paralysis of muscles only on one side. It is very common to have a palsy of one or more branches of the oculomotor nerve in association with paralysis of associated upward or downward movement, and this is more easily explained by a lesion of the oculomotor nucleus than by a cortical lesion. Cortical centers control associated movements, not isolated muscles.

The paralysis of the sixth nerve occurring with paralysis of associated upward or downward movements has been attributed by Lichtheim to pressure upon the nerve at the base of the brain. The opinion held by Parinaud, although supported by Duval, Laborde and Graux, can hardly be accepted, viz., that the sixth nucleus gives nerve fibers to the internal rectus muscle, and that this muscle has a double innervation, the sixth nerve for lateral movement, the third for convergence.

The assumption of a basal coordinating center above the oculomotor nucleus for ocular movements seems forced. The lateral associated ocular movements can be well explained by the connection formed by the posterior longitudinal bundle between the nucleus of the branch to the internal rectus muscle and the nucleus of the sixth nerve. It is reasonable to suppose that certain fibers connecting the different parts of the oculomotor nucleus with one another, or connecting the group of nerve cells innervating the inferior rectus with the nucleus of the trochlear nerve have a coordinating function similar to that of the posterior longitudinal bundle, and that this function may be disturbed by a small lesion. It is striking that the paralysis of downward associated movement (necessitating the implication of two separate nuclei of ocular muscles) without paralysis of upward associated movement is exceedingly rare, far more so than is the isolated paralysis of upward associated movement.

Conjugate deviation of the eyeballs from lesions of the pons is not common when the lesion is confined to one side of the pons. In a case of hemorrhage into the tegmentum of the pons (see Case 3) that I have had the opportunity to study and the brain from which I have examined and refer to in this paper, inward deviation of only the right eye occurred from paralysis of the sixth nerve. One gets the impression that conjugate deviation of the eyeballs away from the side of the lesion is common in lesions of one side of the pons, but in four cases of lesions of this part observed by me (three of tumor and one of hemorrhage) causing paralysis of the external rectus muscle of the same side, I have looked for the conjugate deviation but failed to find it. It is probable that in most cases the side of the pons opposite to the lesion is directly or indirectly involved.

Von Monakow[16] believes that when the lesion implicates the sixth nucleus of one side, apart from the paralysis no disturbance, i.e., no deviation occurs, unless it is the result of secondary contracture. I believe that conjugate deviation is rare even as a late sign of paralysis of one sixth nucleus. Of course any involvement of both sixth nuclei would make conjugate deviation impossible, and in one of my cases (Case 2) both these nuclei were affected.

In a case of softening of the dorsal part of the left side of the pons reported by Albert Ransohoff[17] there was complete paralysis of the associated ocular movement to the left and of convergence movement of the right eye. The right external rectus muscle was not over-active, but the left internal muscle was in marked spasm. The left abducens nucleus was involved in the lesion, the right escaped, but the right posterior longitudinal bundle was involved. Ransohoff mentions that Wernicke and Jolly have shown that paralysis of the external rectus muscle does not in itself cause contraction of its antagonist, the internal rectus, and that in the cases of these authors, as well as in his own, there was a lesion of the opposite posterior longitudinal bundle. In Wernicke's case a tumor had destroyed the left abducens nucleus and invaded the right side of the pons. Both eyeballs were persistently turned to the right, especially the left eyeball.

According to the study of the literature by Ransohoff, contracture of the unparalyzed antagonistic muscles, but not always in both eyes, and more in the internal rectus muscle on the side of the lesion than in the external rectus muscle on the side opposite to the lesion, was observed in the cases of Wernicke, Jolly, Graux, Meyer (the lesion did not extend to the mid-line),

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[16] Gehirnpathologique, p. 355.

[17] Ransohoff, Archiv. für Psychiatrie, Vol. 35, 1901, p. 416.

Spitzer (slight, and after one-half year duration of the lesion), Mierzejewsky and Rosenbach, Gebhard, and Ransohoff.

Ransohoff says that if the posterior longitudinal bundle, as is assumed by most authors, contains a part of the voluntary fibers for the internal rectus muscle of the opposite side, a lesion of these fibers, as of the pyramidal fibers, must cause spastic contracture of the muscle they innervate. Therefore, according to Ransohoff, contracture of the internal rectus muscle on the side of the lesion indicates that the posterior longitudinal bundle of the opposite side is also involved, but this contracture probably could not exist if the nucleus of each abducens were destroyed.

In four of my cases (Cases 1, 2, 3, and Beier) both posterior longitudinal bundles must have been much affected and conjugate deviation did not occur in any.

In one case (Case 4) in which paralysis of lateral movements was present, the movement of either eyeball to the right or left was distinctly greater when one eye was covered than in associated lateral movments. The individual movements of the eyeballs were therefore greater than the associated movements.

One of the cases (Case 9) was especially interesting in that syphilitic infection was positive. The paralysis of associated movement was peculiar; the downward movement was completely lost in the right eye and much impaired in the left; the upward movement was slight in the right eye and completely lost in the left; the lateral associated movements were nearly normal. Almost all the ocular movements were regained except the downward movement of the right eye. It is remarkable that the complete paralysis in the associated movements was in the downward direction in one eye and in the upward direction in the other, although the associated movement was very feeble. It is remarkable that the associated palsy disappeared leaving paralysis of downward movement of the right eye.

In four cases of my series with necropsy (Beier, Cases 1, 2, and 3) the pupil was smaller on the same side as the lesion of the pons. This alteration of the size of the pupil from a lesion of the pons is difficult to explain, but the presence of the sign in four cases shows that it is of diagnostic significance, and also that the fibers controlling the iris pass through the pons.

In at least two cases nystagmus was present in looking away from the side of the chief lesion (Beier, Case 2). This was caused by weakness of the opposite external rectus muscle from pressure upon or direct involvement of the nucleus of the abducent nerve. This sign may be expected when a lesion affects the dorsal part of the pons and destroys the function of one abducent nucleus and weakens that of the other. Nystagmus may surely be the result of paresis of ocular muscles.

I have studied 38 cases of paralysis of upward or downward associated movement reported in the literature, not including my own 9. In many cases certain symptoms are not referred to, so that statements regarding the frequency of symptoms are incomplete. V. Kornilow used only 20 cases of the 27 to which he found references in drawing his conclusions. As a result of my study of these 47 cases.

Paralysis of upward associated movement without paralysis or paresis of downward associated movement was found in 26 cases (Henoch, Priestley Smith (2), Nieden, Gowers, Parinaud (2), Ormerod, Reinhold, Thomsen, Lichtheim (3), Hoesslin, Bruner, Mills, Noguès and Sirol, Kornilow, Posey, Parkinson, cases 1, 2, 4, 5, 6, 8, of my series).

Paralysis of upward associated movement with paralysis or paresis of downward associated movement was found in 16 cases (Wernicke, Parinaud, Hope, Nothnagel, Eisenlohr, Verrey, Sharkey, Sauvineau, Teillais, Gordinier, Raymond and Cestan (slight), Kornilow, Cases 3, 4, 7, 9, of my series).

Paralysis of upward associated movement with impairment of lateral movement, often developing later, was found in 15 cases (Henoch, Priestley Smith (2), Lichtheim (3), Parinaud, Poulard, Raymond and Cestan, Kornilow, Parkinson, Cases 1, 2, 3 and 4 of my series).

Paralysis of upward associated movement without impairment of lateral movement was found in 22 cases (Wernicke, Nieden, Gowers, Parinaud (2), Ormerod, Hope, Thomsen, Sharkey, Sauvineau, Hoesslin, Bruner, Teillais, Gordinier, Noguès and Sirol, Kornilow, Posey, Cases 5, 6, 7, 8, 9 of my series).

Paralysis of downward associated movement without paralysis of upward associated movement was found in 5 cases

(Gee, Poulard, Curzon-Marie and Babinski, Basevi, Schroeder), but in none of my nine cases.

The reaction of the iris was found to be impaired in 14 cases (Henoch, Gowers, Parinaud, Thomsen, Eisenlohr, Lichtheim, Sharkey, Sauvineau, Hoesslin, Gordinier, Mills, Kornilow, Posey, Case 9 of my series) and is said to have been normal in 4 cases (Parinaud, Teillais, Poulard, Raymond and Cestan), but in many cases no statements regarding the iritic reflex are made, and in all my nine cases the iritic reflex was preserved.

The optic nerve was found to be affected in 15 cases (Gowers, Thomsen, Nothnagel, Gee, Eisenlohr, Lichtheim (3), Sharkey, Hoesslin, Gordinier, Cases 1, 5, 7, 9 of my series), and normal in 12 cases (Wernicke, Nieden, Parinaud (2), Ormerod, Reinhold, Sauvineau, Teillais, Raymond and Cestan, Mills, Noguès and Sirol, Posey).

Convergence was impaired in 15 cases (Parinaud (2), Sharkey, Sauvineau, Teillais, Poulard, Gordinier, Raymond and Cestan, Kornilow, Basevi, Cases 2, 4, 5, 8, 9 of my series), and normal, or nearly normal, in 9 cases (Hope, Parinaud, Noguès and Sirol, Kornilow, Posey, Cases 1, 3, 6, 7 of my series.

Ptosis was found in 7 cases (Reinhold, Eisenlohr, Gordinier, Kornilow, Nothnagel, Cases 2 and 8 of my series), and was said to be absent in 13 cases (Wernicke, Parinaud, Gee, Sauvineau, Teillais, Raymond and Cestan, Cases 1, 3, 4, 5, 6, 7, 9 of my series).

Necropsy was obtained in 19 cases (Henoch, Wernicke, Gowers, Hope, Reinhold, Thomsen, Gee, Eisenlohr, Lichtheim (3), Sharkey, Hoesslin, Gordinier, Nothnagle, Basevi, cases 1, 2 and 3 of my series.)

In all of these cases except one (Thomsen[18]) parts about the aqueduct of Sylvius were implicated, and in Thomsen's case a gumma was found in the cerebral peduncles. Thomsen cannot explain why the intense degeneration of the right oculomotor nerve and the slight degeneration of the left oculomotor nerve caused paralysis of only upward movement. It is possible that the Nissl method, if it had been employed, would have shown marked alteration of the oculomotor nuclei, possibly as a toxic effect from the adjacent gumma, and in this way the paralysis of upward associated movement may have been caused. Such was the condition in one of my cases (Case 2), unless perhaps the alteration of the nerve cells was caused by pressure. Until at least another case similar to Thomsen's is reported we may well hesitate to believe that a peripheral lesion of one oculomotor nerve may cause paralysis of only upward associated movements.

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[18] Thomsen. Archiv. für Psychiatrie, Vol. XVIII., 1887, p. 616.

Tumor was found in 14 cases (Henoch, Gowers, Hope, Reinhold, Gee, Lichtheim (3), Sharkey, Hoesslin, Gordinier, Nothnagel, cases 1 and 2 of my series), a bullet wound in one case (Eisenlohr), apoplectic cyst in one case (Wernicke), hemorrhage in one case (my Case 3), and lesion uncertain in one case (Basevi).

Recovery or partial recovery occurred in 7 cases (Priestley Smith, Nieden, Verrey, Kornilow, Cases 5, 8 and 9 of my series). Important symptoms other than ocular palsies were found in 41 cases (Henoch, Wernicke, Priestley Smith (2), Gowers, Parinaud (3), Hope, Reinhold, Thomsen, Nothnagel, Gee, Eisenlohr, Lichtheim (3), Verrey, Sharkey, Sauvineau, Hoesslin, Bruner, Teillais, Poulard, Gordinier, Cruzon-Marie and Babinski, Raymond and Cestan, Mills, Noguès and Sirol, Kornilow (2), Schroeder, Posey, Parkinson, Cases 1, 2, 3, 4, 6, 8 and 9 of my series).

As a result of my studies I believe that persisting paralysis of associated lateral movement indicates a lesion of the posterior longitudinal bundle; that persistent paralysis of associated upward or downward movement indicates a lesion in the vicinity of the oculomotor nucleus, and that paralysis of associated ocular movements is not the result of a lesion of extracerebral nerve fibers. Lesions of the cerebral cortex may certainly cause paralysis of lateral associated ocular movements, and possibly of upward or downward associated ocular movements, but cortical paralysis of associated ocular movements is transitory, unless possibly where the center on each side of the brain is destroyed. Paralysis of associated ocular movements may be caused by hysteria. Any case in which associated ocular palsy is persistent and is of organic nature, is unsuitable for operation unless the operation is merely palliative, as the lesion is probably within the posterior part of the pons or cerebral peduncle, according to the form of the associated palsy, or else causes much pressure upon the dorsal portions of these structures. The paralysis of associated ocular muscles may be produced by inflammatory lesions or lesions of a similar character (alcohol, syphilis) as well as by tumor, and may disappear later in the course of the disease. Syphilitic ependymitis and cellular infiltration must always be considered in diagnosticating the lesion causing paralysis of associated ocular movements.

CASES OF PARALYSIS OF ASSOCIATED UPWARD OR DOWNWARD MOVEMENT OBSERVED BY DR. SPILLER.

Case I. Tubercle of left side of the pons.

Galik, was admitted to my wards in the Philadelphia General Hospital, Aug. 11, 1904, and died Oct. 22, 1904.

He said he had been paralyzed for nine months on the right side of the body. He was a fairly well developed man, aged thirty-one years. He was examined Aug. 12 by Dr. T. H. Weisenburg, when the following notes were made:

He never had headache nor vertigo, and has had no difficulty in chewing or swallowing, this statement being corroborated by the nurse. There is no dribbling of saliva. He has internal deviation of both eyes, more of the left. The right pupil is larger than normal, the left is normal in size. Both pupils respond to light and in accommodation and convergence. The left external rectus is completely paralyzed, and the right external rectus is partially paralyzed. All of the other movements are well performed. On showing his teeth he can draw back the right angle of his mouth better than the left. The right nasolabial fold is more distinct than the left. On attempting to close his eyes the left lids remain partially separated. He wrinkles the right side of the forehead normally, but there is hardly any movement on the left side. The tongue deviates to the right when protruded, but is not atrophied and shows no tremor. The masseter muscle does not contract so well on the left side as on the right.

He is able to lift the right upper limb from the bed for a few minutes, but the movement is unsteady and weak. Resistance to passive movement at the right elbow is almost lost. The right upper limb seems somewhat spastic. The right forearm is held in supination, the hand flexed on the wrist, and the fingers flexed into the palm. The grip is very feeble. There are no contractures, but the fingers are stiff. The whole right upper limb is wasted, more so in the distal parts, the muscles of the thenar and hypothenar eminences being almost completely atrophied. Atrophy of the extensors and flexors of the forearm is more marked on the right side than it is in the corresponding muscles of the left upper limb. The biceps, triceps and wrist reflexes on the right side are prompter than normal.

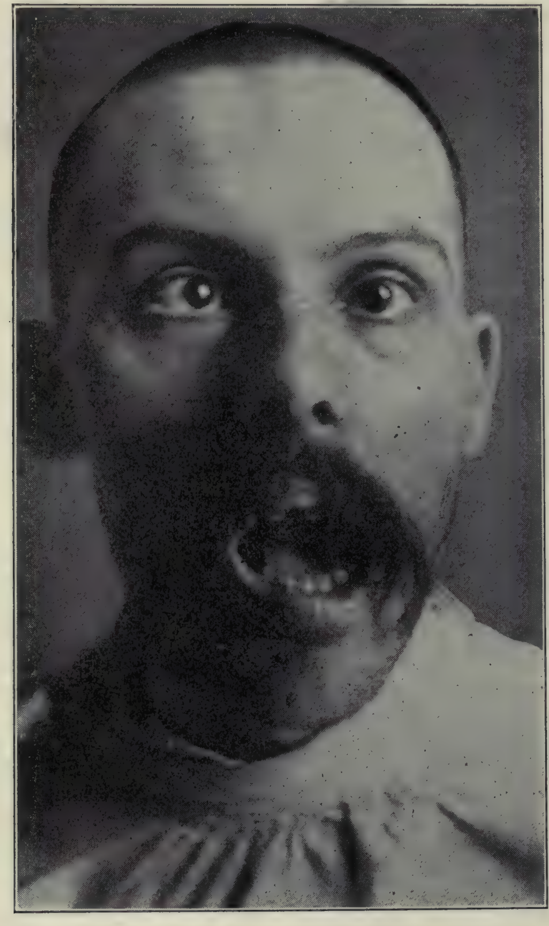

FIG. 1. Internal deviation of both eyes, especially of the left, and deviation of the lower jaw to the left from paralysis of the left muscles of mastication. Paresis of associated movements upward and to the left.

Sensation for touch is felt in the right upper limb, right side of chest and abdomen, but less so than in the left side. He does not recognize a pin prick over these parts on the right side as a pin prick, but as touch, and sensation is more impaired in the distal parts. The left upper limb is normal in every respect. He is not able to recognize objects placed in his right hand.

The power of the right lower limb is diminished. He is able to flex and extend the thigh, but is not able to move the ankle joint or the toes. The motor power in the left lower limb is normal. The patellar and Achilles tendon reflexes are much increased on the right side, but not on the left. Sensations for touch and pain are diminished in the right lower limb, and more so in the distal parts; they are normal in the left lower limb. The right lower limb is somewhat spastic and atrophied, and the atrophy is seen especially in the thigh muscles. He has had no bladder or rectal disturbances.

He says he has no pain anywhere.

His speech is thick and indistinct.

Dr. Van Epps saw the patient Aug. 13, and made the following notes: Contraction of the left orbicularis palpebrarum is markedly weakened, that of right is questionably weakened. Right pupil is round, 3 mm. in size. Left pupil is round, 2 mm. in size. Both pupils react to light and in accommodation and convergence. Outward rotation of O. D. 2 mm. short; internal rotation 3 mm. short, and upward 2 mm. short of full; downward rotation normal.

O. S. inward rotation normal, complete absence of outward rotation, upward and downward rotation similar to that of fellow. On this date some weakness of the right orbicularis palpebrarum was noticed. The nurse said the patient had difficulty in swallowing.

Dr. Van Epps made a further report Aug. 16, 1904.

O. D. Under homatropine, pupil 6 mm. round; media clear, marked optic neuritis with swelling of retina for 1½, disk clean in all directions (neuroretinitis). Disk elevation 2 diopters. Arteries reduced in size, veins slightly increased in caliber and quite tortuous, partly disappearing here and there in swollen nerve and retina. Hemorrhage on upper part of disk. Middle and peripheral zones normal. O. S. Under homatropine 4½ mm., round. Cornea epithelium so eroded that a view of the fundus is impossible.

Entire loss of corneal and conjunctional reflex on the left side (hemianesthesia of same complete).

There seems to be marked hyperesthesia of eyelids and surrounding cutaneous area on the left side, but patient's answers are contradictory.

He was seen by me Aug. 25, at which time I made the following notes:

In converging the movement of the right eyeball inward is about equal to that of the left, and is greater than when the attempt is made to look toward the left with both eyeballs. He has paralysis of associated movements, both in looking to the left and upward. The movement of each eyeball upward is impaired equally. When he closes his eyelids he has marked fibrillary tremor of the left upper lid. He has marked fibrillary tremors of the left masseter and the left masseter is completely paralyzed. When the mouth is opened the jaw goes markedly toward the left. Speech is distinctly bulbar. Hearing cannot be tested carefully on account of slow cerebration, but seems to be affected on the left side. The weakness of the right upper and lower limbs is very pronounced, though it is not complete paralysis. The tendon reflexes of the right upper and lower limbs are distinctly exaggerated.

He is able to stand, but not without support.

He swallows with difficulty.

The left cornea is anesthetic.

He says pin prick is felt more in the left side of the face. He draws up the corner of the mouth when the corresponding side of the face is stuck with a pin. This sensory condition was noticed also by Dr. Weisenburg, who found (Aug. 12, 1904) that he recognized touch and pain sensations distinctly better on the left side of the face than on the right.

Sept. 21. Speech is very indistinct and is bulbar in character. The left side of the face is paralyzed. Marked fibrillary tremors are seen in both sides of the tongue, and the tongue deviates to the right when protruded. The left pupil does not reach the median line, and the right pupil goes a little beyond the median line in trying to look to the left. Pain and tactile sensations are diminished in the right upper limb. Babinski's sign is absent on each side. Ankle clonus is not present on either side.

Sept. 23. He does not hear a low ticking watch when placed on the left ear, but does hear it when it is placed on the right ear. He has excessive appetite.

Sept. 28. The left eye is much congested, although mosquito netting is kept constantly over his face to keep the flies away and his eyes are washed frequently with boric acid. Pus is found in the anterior chamber of the eye.

Oct. 22. He has been gradually sinking more than ten days. Respirations have become shallow and rapid, and he is unable to speak. He died at 8.30 p.m., Oct. 22, 1904.

Death occurred while the patient was in the service of Dr. C. K. Mills, after I had gone off duty. I am indebted to Dr. Mills for the pathological material.

At the necropsy ulcerative tuberculosis of the lungs and a large tumor in the left side of the pons were found.

Sections made through the cerebral peduncles show that the aqueduct of Sylvius is compressed on the left side. The left cerebral peduncle is considerably large than the right. The tumor extends into the lower part of the left cerebral peduncle. The left fourth nucleus appears to be almost normal, a few of the cells are slightly altered, but the nucleus on the whole is remarkably well preserved. The posterior longitudinal bundle on the left side is pushed more dorsally than is the corresponding bundle on the right side. The right 4th nucleus also is normal except that a few of the cells show alteration. Most of the nerve cells in the oculomotor nucleus, both in each lateral and in the median group seem to be normal, only occasionally a cell can be found in which the chromophilic elements have undergone disintegration.

Left 2nd nerve. There is a slight round cell infiltration of the pia and of the septa extending into the optic nerve; the nerve does not appear to be degenerated in its intracranial portion.

Left 3d nerve appears to be entirely normal.

Left 6th nerve is much degenerated. The medullary sheaths are swollen. The axis cylinders are also swollen and in some places have disappeared. One side of the nerve is much more altered than the other. The Weigert stain shows much degeneration at one side of the nerve where the medullary sheaths stain very badly. The right sixth nerve appears to be normal.

Left 7th nerve shows a moderate swelling of medullary sheaths.

Left 8th nerve appears to be normal.

Left 5th nerve is almost completely degenerated in a few bundles that probably belong to the motor part of the nerve.

The left pyramid is much degenerated by the method of Marchi, as is also the spinal root of the left fifth nerve. There are some degenerated fibers on each side of the medulla oblongata posterior to the lower olive, more on the left side, and also in the left lemniscus.

The right crossed pyramidal tract is much degenerated by the Marchi method from the cervical into the lumbar region. The left direct pyramidal tract is much degenerated in the cervical region.

There is no meningitis about the medulla oblongata or spinal cord. The nerve cells of the anterior horns of the cervical and lumbar regions stain well by the thionin method.

Weigert hematoxylin sections show intense degeneration of the left anterior pyramid, of the left direct pyramidal tract, and of the right crossed pyramidal tract.

The tumor in the middle portion of the pons, antero-posteriorly, invades a little the right side of the pons (see Fig. 2), but the greater part of the right pyramidal tract is not involved in the growth. The aqueduct of Sylvius is much compressed and appears as a long narrow slit. Almost the entire left half of the pons is destroyed, a narrow zone at the ventral part escaping. The tumor in transverse section is nearly round. Microscopically the tumor is not sharply defined from the surrounding tissue, and its peripheral zones consist of mononuclear cells with round nuclei. The central portion of the tumor is degenerated, and the cells do not stain well. The tubercle bacillus was not found. The tumor has the appearance of a tubercle.

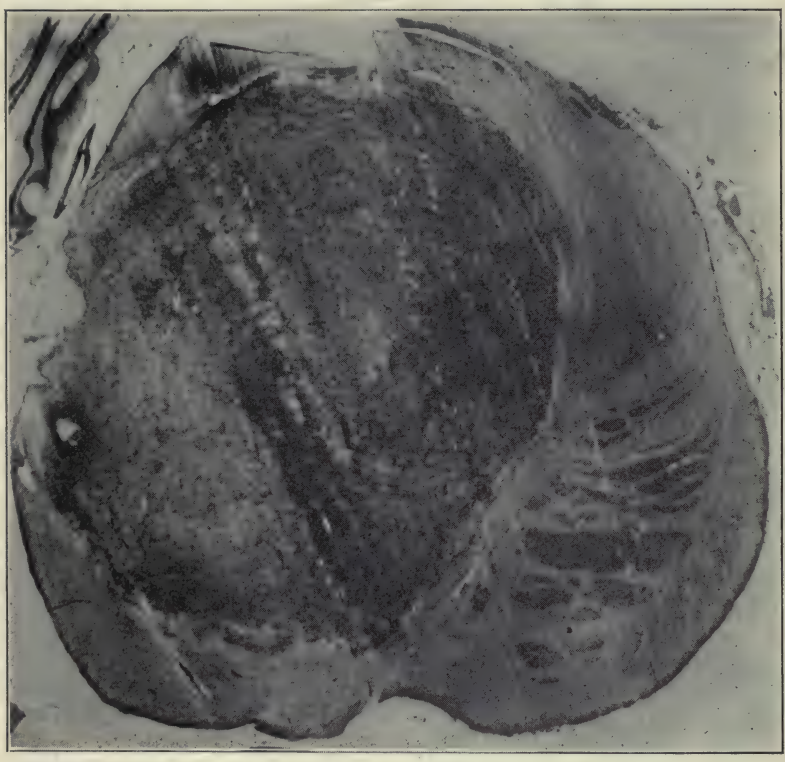

Fig. 2. Tubercle in the left side of the pons.

Summary: B. Galik, a male aged thirty-one years, had been paretic (Aug. 11, 1904) nine months in the right limbs. He was paretic in both sides of his face, especially in the left side, and in the left muscles of mastication, and had fibrillary tremors of the left masseter muscle, and of the right side of the tongue. The tendon reflexes in the right upper and lower limbs were exaggerated. Diminution of sensation and some wasting were observed in the right upper and lower limbs. The movements of both eyeballs upward were impaired, and there was complete loss of movement of the left eyeball to the left, and marked impairment of movement of right eyeball to the left; also, impairment of movement of right eyeball, but not of left eyeball, to the right. Downward movement of both eyeballs was normal, and convergence was preserved (paresis of associated movements upward and to the left). He had bulbar speech, impaired hearing in the left ear, anesthesia of the left cornea, optic neuritis, at least in the right eye (left disc not visible because of corneal opacities); and later left neuroparalytic keratitis.

The necropsy showed a large tubercle involving almost the entire left side of the pons and invading a little the right half of the pons and extending into the lower part of the left cerebral peduncle. Most of the nerve cells of the oculomotor nuclei appeared to be normal, but those of the left nucleus were displaced. The left 3d nerve was normal. The left sixth nerve and spinal root of the left 5th nerve were much degenerated, and the left 7th and left motor 5th nerves were partially degenerated. The left pyramid was degenerated. The aqueduct of Sylvius was much compressed from the left side.

The slight voluntary movements of the right upper and lower limbs may be explained by the preservation of a narrow zone in the ventral portion of the left half of the pons and by axis cylinders that probably passed through the tubercle without being destroyed. It has been shown that axis cylinders may be found within a tubercle even after the medullary sheaths have been destroyed.

The paralysis of associated left lateral movement is to be explained by the implication of the left posterior longitudinal bundle. The paresis of upward movement could not have been caused by implication of the corpora quadrigemina, because this portion of the brain stem was not involved. The nerve cells of the oculomotor nuclei were slightly degenerated, but the left oculomotor nucleus was much displaced in its posterior portion, and the function of the nerve cells here must have been interfered with.

The pupil in this case was smaller on the side of the tumor.

It seems difficult to explain the absence of paralysis of associated ocular movement to the right; unless it is because the ocular examination was made two months before death occurred, and at that time the turbercle had not extended to the right side of the pons. The man's intelligence became considerably affected before death.

Case 2. Glioma of the right side of the pons.

A. Moore, male, aged sixteen years, was a patient of Dr. Wm. Evans, from whom the following notes were obtained:

His parents and two sisters were living and well. His maternal grandfather died of apoplexy at seventy-one years. He had always had good health until the early part of July, 1904, when he began to feel nervous, slight exertion made him tired, he became dizzy, had headache and a tendency to turn to the left, and he staggered slightly in walking. His sight became dim, and at times he had diplopia.

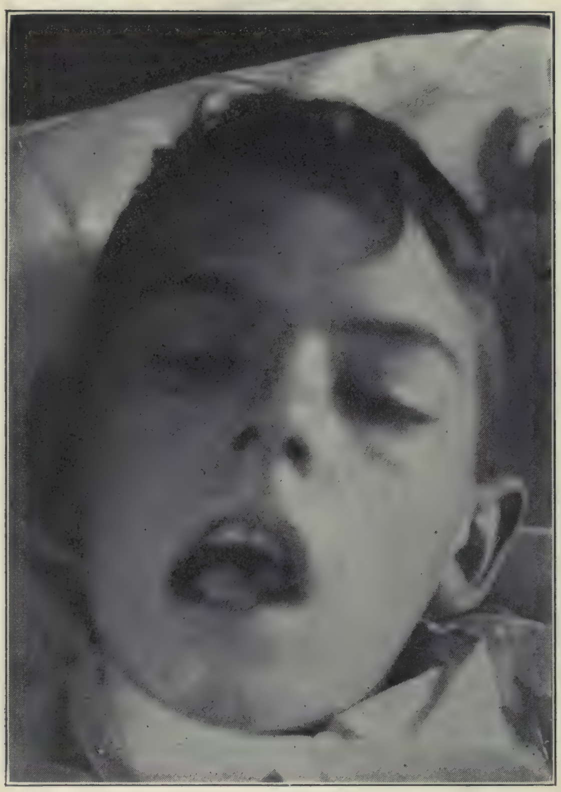

FIG. 3. Attempt to converge. Notice position of left eye. The left eye is moved more toward the right than when the attempt is made to move both eyeballs to the right (see Fig. 5). Notice the marked deviation of the lower jaw to the right when the mouth is opened, this being the result of paralysis of the right external pterygoid muscle.

He was seen by Dr. C. S. Potts Aug. 15, 1904, and notes made by him at that time are as follows:

He sits with the head retracted and bent to the left, the chin pointing to the right, although he can hold it straight if he makes an effort to do so. He has ptosis of the left eyelid. The pupils are equal and respond to light and in accommodation. Some weakness of the external rectus of the right side is detected and he says that he sees double when he looks to the right. Coarse nystagmic movements are present when the eyes are moved laterally in either direction. The angle of

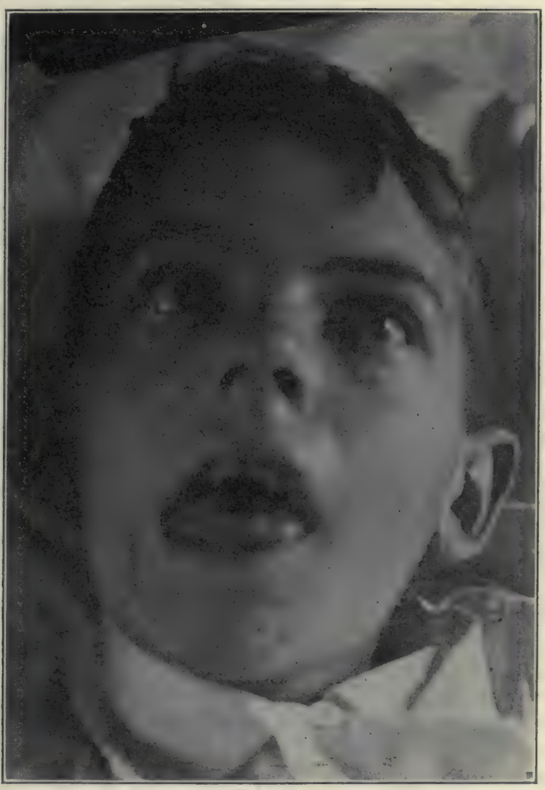

Fig. 4. Attempt to look upward and to protrude the tongue. The eyeballs move very slightly upward. The lower jaw goes to the right, the tongue to the left when protruded. The dot on tongue shows the point of the tongue. On great effort, the tongue could be protruded a little further, and the deviation to the left was greater.

the mouth droops slightly on the right side, but there is no distinct paralysis. The tongue seems to protrude slightly to the left. The gait is staggering, but there is no tendency to fall to one side more than the other. Resistance to passive movements is good. The patellar reflexes are exaggerated, the left more than the right, and he has slight ankle clonus on the right side and marked clonus on the left side. Plantar reflexes are diminished, the toes move in neither direction on the right side, but slightly upward on the left side. The arm jerks (biceps, triceps, wrist) are apparently equal on both sides and about normal in degree. He hears the tick of a watch in the right ear at 12 inches, in the left ear at 3 feet. Bone conduction is better on the left side. He can recognize the direction of passive movements of the fingers and toes, but cannot touch the end of his nose with either index finger with certainty. He has not other sensory symptoms except headache.

An examination of the eyes by Dr. Krall, Aug. 17, 1904, gave the following results:

He has partial ptosis of left eye, paralysis of the left external rectus, or rather a paresis, paresis of the left inferior rectus, and a nystagmus to the right on attempting fixation. Accommodation for his age is good. The pupils are nearly alike on the average (the left one is ½ mm. larger than the right), they react normally to light, accommodation and convergence. His vision is practically normal. The ophthalmoscopic examination shows no involvement of the optic nerve. Field of vision is, however, contracted concentrically for form and color. Color perception is good. No color blindness.

The patient was seen by me Oct. 11, 1904, in consultation with Dr. Evans, and at that time I made the following notes:

The left pupil is considerably larger than the right, Iritic reflex is prompt in each eye. Nystagmoid movement in trying to look to the left. Movement of eyeballs to the left imperfect. Left iris does not reach the external canthus and the eyeball returns almost immediately towards the median line. Unable to look towards the right with either eye, pupils remaining in the median line when he attempts to look to the right. Able to converge very slightly with the left eye, and does so with the right. Upward movement of each eye is almost lost, the pupils hardly passing beyond a line drawn horizontally across the centers of the palpebral fissures. Downward movement seems almost normal. He closes the eyelids firmly on each side. Slight drooping of each lid is present. He wrinkles the forehead well and equally on both sides. The movements of the face are feeble on each side in the lower distribution of the seventh nerve. He does not show the teeth well nor does he draw up either corner of the mouth well. Tongue when protruded goes distinctly towards the left. Left masseter muscle contracts, the right apparently does not. When the mouth is opened the lower jaw goes far to the right. The right orbicularis palpebrarum is distinctly weaker than the left, and often in closing his lids, the right lids remain partially open. Pin prick is promptly perceived on each side of the face. It is probable that the right side of the face in the lower distribution of the facial nerve is a little weaker than the left side. The neck is not stiff. There is a tendency for the head to turn to the left, but it can be turned easily to the right. He hears a low-ticking watch 6 to 8 inches in the left ear. Cannot hear it in the right ear when placed against the ear. Speech is thick, bulbar in type, not at all aphasic. He counts fingers correctly

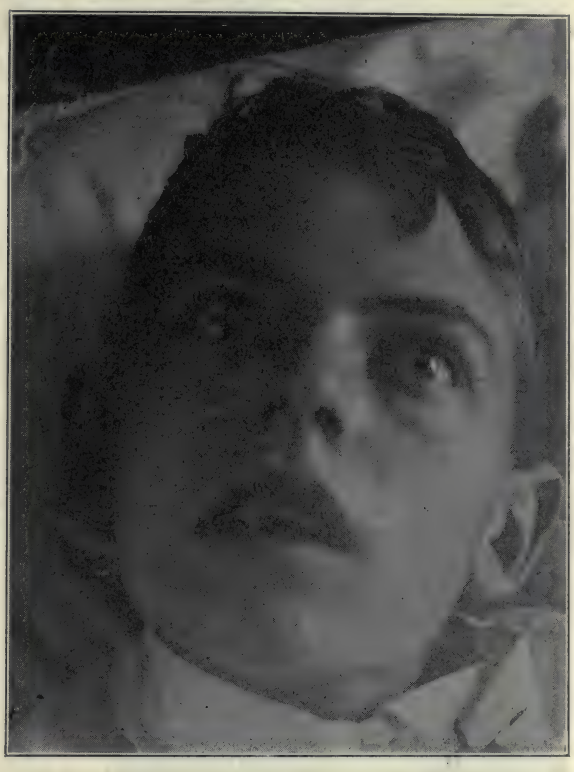

FIG. 5. Attempt to look to the right and to separate the lips with the teeth together, showing weakness in the lower distribution of the facial nerve on each side. Notice the position of the left eye in this photograph and in Fig. 3, showing attempted convergence.

held before either eye. He is able to swallow liquids or semisolids without much trouble. The soft palate moves freely on voluntary effort. Finger-to-nose test shows extreme ataxia with right hand. The left upper limb is almost completely paralyzed, but there is slight movement of the left shoulder. No motor power is preserved below the shoulder in the left upper limb. Biceps and triceps reflexes are slightly increased on the left side, almost normal on the right side. Sensations for touch and pain are somewhat diminished in the left upper limb, and are normal in the right upper limb. Sense of position seems to be preserved in the left upper limb, as he puts his right hand directly on his left with eyes closed, when told to do so. Upper

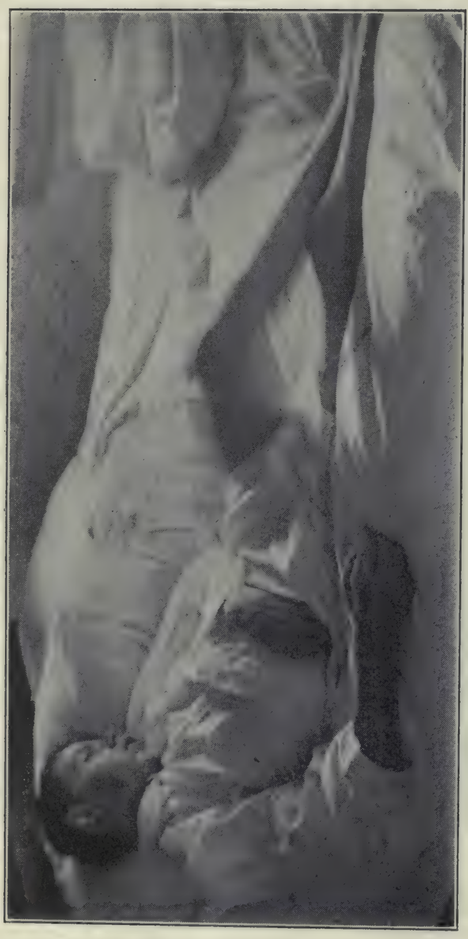

Fig. 6. Showing the tendency of the head to fall forward and to the left; also the complete paralysis of the left upper limb and the partial voluntary movement of the left lower limb at the hip. The voluntary power at the left shoulder was very slight.

limbs are not distinctly wasted. Left lower limb is distinctly weaker than the right, but not as weak as the left upper limb. The right lower limb has normal power. The lower limbs are not distinctly wasted although the whole body is somewhat emaciated. The patellar reflex is slightly increased on the right side, distinctly so on the left. Tendency to patellar clonus is present on the left side only. Persistent ankle clonus on both sides, but much more distinct on the left side. Typical Babinski sign is obtained on the left side, but not on the right side, there being no movement of the toes at all on this side. Sensations for touch and pain are probably diminished slightly in left lower limb. Heel to knee test shows much ataxia of right lower limb. When sitting on the edge of the bed he would fall if not supported, and head falls far forward, and to the left. When made to stand he would fall if not supported. He cannot stand erect, and ataxia is so great that he does not seem to know where to put his legs. His knees flex under him. He understands all that is said to him. Sitting up makes him dizzy. He has ocasional attacks of dull headache in the right temporal region. The tongue is not wasted, nor does it show fibrillary tremor. Almost every day he has attacks of nausea, sometimes with vomiting.

In converging left eyeball moves distinctly a little more toward the right than when he tries to look to the right with both eyeballs.

Further notes were made by me Oct. 23, 1904.

Upward movements of eyeballs much impaired, but seem to be better than when he was last seen. The jaw goes far to the right. Tongue not protruded much beyond the lips, but deviation to the left occurs. Movement of eyeballs to the right is lost, much impaired toward left and nystagmus occurs when eyes are turned toward the left. Weakness of right orbicularis palpebrarum is more pronounced. Right eyeball moves up a little more than does the left. He raises the left eyebrow better than the right. He does not hear a loud-ticking watch unless placed directly on the right ear, but hears it a foot away from the left ear. Sensations for touch and pain are not much impaired on either side of the face. He certainly feels touch and pain in the left forearm. The movement in the left lower limb is almost entirely at the hip. He feels both touch and pain sensation in the left lower limb. He has no movement in the left toes or at the left ankle. When he sits up the head falls far forward and to the left and the whole body inclines forward.

The necropsy was obtained Nov. 10, 1904.

The left 4th nucleus is partly degenerated, some of the nerve cells of this nucleus show much degeneration, but most of them are in excellent condition. There is no distinct group of cells constituting the right 4th nucleus at this level, because the right 4th nucleus has been pushed more cerebralwards by the tumor. The right posterior longitudinal bundle has been pushed dorsally toward the aqueduct of Sylvius, and the aqueduct is distorted in such a way that its right half is pushed further dorsally than its left half. The glioma extends into the right cerebral peduncle as high as the level of the fourth nucleus, but does not invade this nucleus. Some of the cells of the right fourth nucleus are degenerated, but most of them are normal.

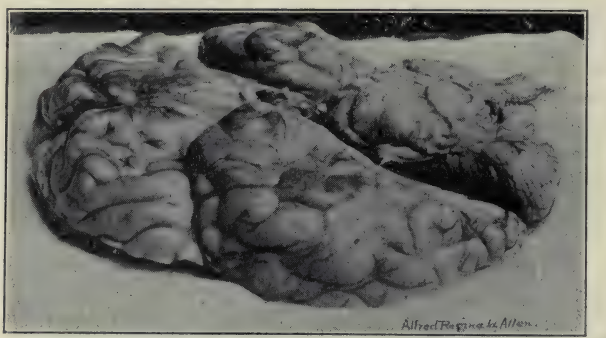

Fig. 7. Glioma of the right half of the pons, invading the lower left half of the pons.

The median group of cells in the oculomotor nucleus is intensely altered, the chromophilic elements have become fine granules and the nuclei are displaced far to the periphery of the nerve cells. The left lateral group in the oculomotor nucleus is well developed, although here and there a nerve cell is found that is not quite normal. The right lateral group shows much alteration, and yet not nearly so much as the median group. The nerve cells are much less numerous in the right lateral group than in the left lateral group, and they are pushed backward toward the aqueduct of Sylvius; many of them appear to be normal. Cells bordering on the upper and median side of the right lateral group are intensely degenerated. The right oculomotor nerve in its extrapeduncular portion does not appear to be degenerated.

The right second, fifth and sixth nerves and the left sixth nerve do not appear to be degenerated either by the Weigert hematoxylin or acid fuchsine method.

The right half of the medulla oblongata is much larger than the left, but does not contain any tumor tissue at the level of the twelfth nuclei. The twelfth nuclei and the intramedullary part of the twelfth nerves appear to be normal. The right anterior pyramid takes the Weigert hematoxylin stain a little less deeply than the left, but the degeneration is slight.

Sections from the medulla oblongata stained by the Marchi method show some recent degeneration of the right anterior pyramid, and of each column of Burdach, but not of the columns of Goll.

Sections from the uppermost part of the cervical cord show considerable recent degeneration of the left crossed pyramidal tract, and of the columns of Burdach, and slight recent degeneration of the right crossed pyramidal tract.

The comparatively slight degeneration of the pyramidal tract from the right side of the brain is striking in consideration of the size of the tumor.

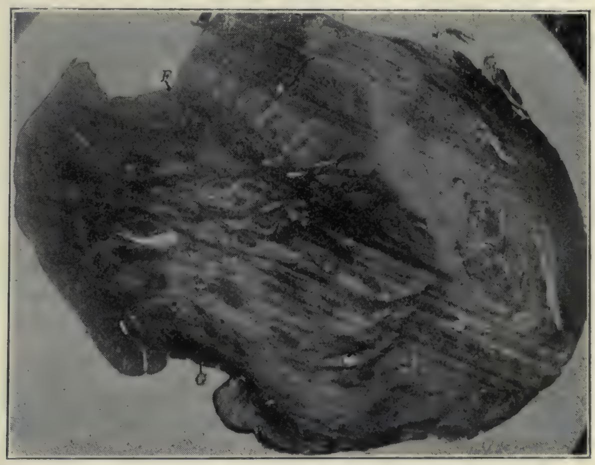

Fig. 8. Microscopical section through the upper part of the pons, showing the glioma of the right half of the pons. FG=raphe of pons.

A section made through the upper part of the pons shows that the tumor is entirely in the right half of the pons, does not reach the raphe, and leaves intact many bundles of the pyramidal tract on the right side, although they are more widely separated from one another than is normal. The right lemniscus is not degenerated. The tumor is chiefly in the lateral part of the right side of the pons, and here is cystic. The tumor consists of cells containing small round or slightly oval nuclei and are separated from one another by much intercellular fibrous tissue. The tumor is a glioma, and in some places is very vascular. It invades the left side of the pons nearer the medulla oblongata.

Summary: A. Moore, male, aged fifteen years, (Oct. 11, 1904) had been complaining about nine weeks. The left pupil was larger than the right. Iritic reflexes prompt. The movements of the eyeballs to the left were imperfect, and to the right lost. Upward movement was much impaired, downward movement about normal. Convergence preserved, but impaired. (Paralysis of upward and lateral associated movements.) Slight bilateral ptosis. Weakness of each facial nerve and of the left side of the tongue. Paralysis of the right masseter muscle and of the left limbs. Tendency to fall to the left. Impairment of hearing in the right ear. Bulbar speech. Diminution of sensation in the left limbs.

A large glioma was found in the right half of the pons, extending into the right cerebral peduncle and the right side of the medulla oblongata, and in the lower part of the pons involving the left side of the tegmentum of the pons. The nerve cells of the 4th and 3d nuclei were partly degenerated, especially on the right side. The left crossed pyramidal tract showed considerable recent degeneration.

The slight impairment of sensation may be explained by the slight involvement of the lemniscus, but it is remarkable that the left motor paralysis was so intense when so much of the motor tract in the right side of the pons escaped. In the left upper limb the only voluntary movement was slight drawing up of the shoulder, and in the left lower limb slight flexion at hip and knee.

Case 3. Hemorrhage of right tegmentum of the pons.

F. D., aged forty-four years, stone mason, was admitted Feb. 8, 1902, to the service of Dr. Arthur V. Meigs in the Pennsylvania Hospital.

Family history is negative.

Previous history: He had been drinking much for a month before his symptoms developed. Yesterday (Feb. 7) was at work cutting stone and feeling well, when he had a stroke at 4 p.m. He fell to the ground but did not lose consciousness and had strength enough to crawl to a block of stone and sit down. His friends noticed his loss of power on the left side and peculiar inward squint of right eye, which, however, disappeared before he reached the hospital, but returned the following day. He had no real convulsions but shook all over after the stroke.

Present condition: He is a short, chunky Italian and cannot talk plainly, and has loss of power on the right side of the face. Pupils are equal and much contracted. Eyes are straight. Beads of sweat are on the left half of face, which retains the normal wrinkles, while the right side has lost all of these. The tongue can be but slightly protruded, and deviates slightly to the right, and is not tremulous. Respiration is good and even. Fremitus, percussion note, and breath sounds are normal both front and back.

Heart: Pulse is strong and full. Apex beat is not visible. Dulness extends from the left border of sternum to within 2 cm. of nipple line. There are no murmurs.

Liver: Dulness from 5th rib to costal margin in nipple line.

Spleen: 9th to 11th rib and is not palpable.

Abdomen fat and soft.

He has only partial power of the left upper and lower limbs. Ankle clonus is not present. The left patellar reflex is increased, the right is normal. He has no pain and lies in comfort, inconvenienced only by loss of power.

Urine is acid, 1020, trace of albumin, no sugar, many hyaline and granular casts, uric acid crystals.

The patient was seen by me Feb. 17, 1903, at which time I dictated the following notes:

Left pupil is considerably larger than the right and the left eyeball protrudes a little more than the right, and the left palpebral fissure is a little wider than the right. Each pupil contracts promptly to light but soon dilates again. Contraction of the pupils is probably prompt in convergence. Accommodation reaction can not be tested. He cannot close the right eyelids. In looking upward the forehead is not wrinkled on the right side. In showing his teeth only the left corner of the mouth is drawn upward. He has therefore complete or almost complete paralysis in the entire distribution of the right 7th nerve. Cannot protrude the tongue beyond the lips, and it deviates slightly toward the right.

Masseter muscle contracts well on the left side, but much less strongly on the right side, and when the mouth is opened the jaw goes a little to the right. He has therefore weakness of the right masseter and right pterygoids. Apparently he has immobility of the eyeballs, as he is unable to look in any direction other than straight forward. His intelligence is feeble but is sufficient to enable him to understand simple questions.

When his face is stuck with a pin on either side he does not seem to have great pain. If anything, he gives more evidence of pain when he is stuck on the right side.

Voluntary power in the right upper limb is good, while the left is paretic but not completely paralyzed. While he can move the whole upper limb, he cannot raise it above his head or close the fingers firmly.

Tendon reflexes of each upper limb: Biceps and triceps tendon reflexes are not very distinct, and are not greater on the left side than on the right. He gives very little evidence of pain when either upper limb is stuck with a pin. He is weak in the left lower limb, but has still some movement in this limb. He can draw this limb up slightly at the hip and knee, and can move the toes. The movements of the right leg are much better than those of the left, but probably are not fully normal. The patellar reflex is diminished on each side, but is better on the right side. Ankle clonus is not present on either side. The Achilles jerk is diminished on each side. The Babinski reflex is not obtained on the left side, but the toes are neither flexed nor extended. On the right side the movement of the toes is distinctly flexion. Sensation of pain is probably preserved in each lower limb.

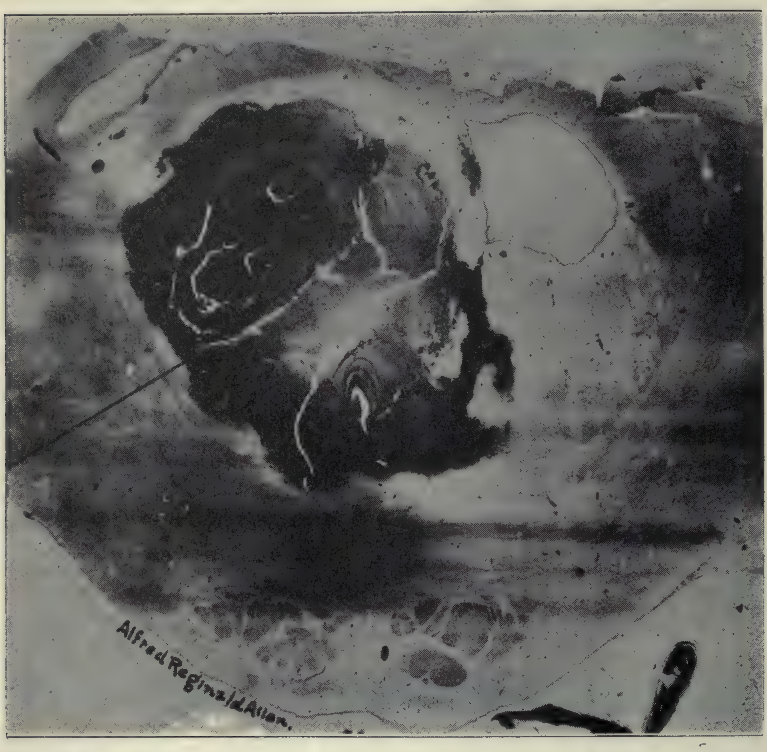

FIG. 9. Line shows hemorrhage of the right tegmentum of pons. Inward deviation of the right eyeball. Immobility of both eyeballs ten days after the apoplexy. Temporary paralysis of all associated movements. Convergence preserved, although weak.

On attempting to converge the right eyeball moves down and inward, but the left eyeball moves only slightly downward.

Feb. 20. He has been quiet. Bowel movements have all been involuntary. Temperature has gone up somewhat and breathing is difficult. He has a tendency to let the tongue fall back and obstruct the ingress of air. Hands are twitching at times. Left pupil is larger than the right. Inward squint of right eye continues. Immobility in lateral direction continues.

Feb. 22. Condition while very alarming yesterday is now much improved. Breathing is easier, intelligence is better. He recognized and talked with his son.

Feb. 24. The eyes have regained a large amount of lateral movement [the direction not stated]. Hand grasp slightly firmer, pulse moderately good. Temperature touched normal yesterday.

Feb. 26. Condition gradually getting worse, but no special change in nervous symptoms. He is weaker.

Temperature during first few days after admission was between 99 and 101 degrees, then dropped below normal for several days, but on Feb. 20 rose again and remained more or less elevated until death.

Feb. 28. Patient died at 1.30 a.m.

The clinical diagnosis I made in this case was hemorrhage of the right tegmentum of the pons, and it was confirmed by the necropsy.

Important in this case were the following symptoms developing after an apoplexy: Contraction of right pupil, almost complete paralysis in the entire distribution of the right facial nerve, paresis of the right side of the tongue, paresis of the right muscles of mastication, weakness of left upper and lower limbs, but not paralysis, inward deviation of the right eyeball, immobility of both eyeballs ten days after the apoplexy. There was therefore at least temporary paralysis of all associated movements. Convergence was preserved though probably was not normal. It is not probable that the paralysis of upward and downward associated movements would have been persistent if the patient had lived, although the paralysis of right lateral associated movement would probably have remained.

Case 4. This case has been reported by Dr. William Campbell Posey[19] when the disease was in an earlier stage.

The early notes are taken from Dr. Posey's paper.

"F. S., blacksmith, forty-eight years of age, came to the Wills Eye Hospital upon Sept. 22 of this year, on account of failing sight. He had been in attendance at the clinic a year previously on account of supraorbital headaches and general presbyopic symptoms, and had obtained a pair of glasses which gave him normal reading power in both eyes. The examination at that time showed normal fundi; the pupils were noted as being 3½ mm. in size, and the irides responded promptly to light, but sluggishly upon convergence. No note was made of any limitation in the extraocular motions or of any unusual position of the head.

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[19] Annals of Ophthalmology, July 1, 1904.

"Corrected vision was normal in both eyes. The patient continued his trade until 3 months ago, when he was compelled to desist on account of attacks of vertigo. These attacks would come on suddenly, without premonitory signs, and were usually induced by change of posture. He had no headache, neither nausea nor vomiting, and his body weight was well maintained. He was mentally depressed, however, most of the

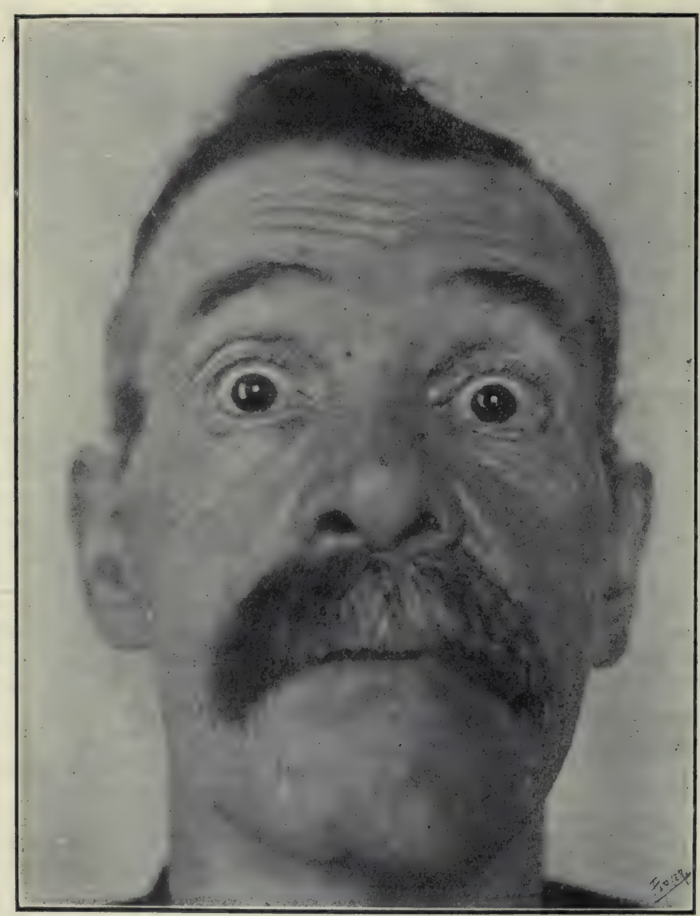

FIG. 10. Paralysis of upward movements of the eyes showing widening of the palpebral fissures and lagging behind of the eyeballs when patient endeavors to look up. (Photograph from Dr. Posey's paper.)

time. The patient denies specific infection, but acknowledges masturbation. During 25 years he drank very heavily, being practically in a continual state of intoxication the most of that time; for the past 5 years, however, he avers that he has drunk nothing. He says that he never chewed, but that he has smoked about ½ package of tobacco a week since he was a boy. Although subject to slight rheumatic pain, he has never had a severe attack of rheumatism or of any other systemic disease. He has never suffered a traumatism or a fall. His family history is negative.

"While sitting quietly upon the bench, waiting his turn to present himself for examination, the writer noted that the patient presented a peculiar fixity of gaze, which was found to be occasioned bv the fact that the eyes were kept fixed in the primary position and that the patient moved them but little to right or left but sat with his gaze directed straight in front of him. The patient looked anxious and changed the expression of his face but rarely. Closer inspection revealed that the head was thrown slightly back and that several mm. of clear sclera intervened between the margin of the upper lids and the upper limbus of both corneas. When requested to fix the examining finger held in front of the eyes and in the median line, the head was thrown further back and, as the finger was carried upward this was more and more marked, while the eyes remained fixed, not rotating above the horizontal plane. The palpebral fissures grew wider as the patient endeavored to look up (see accompanying photograph) and as the eyes lagged behind, the impression was given of an inverted Graefe lid sign. Lateral motions to the right and left were normal, though the eyes followed the movements of the finger somewhat tardily. Downward motion, too, was conserved, though rotation in this direction was accomplished with an effort and only after several trials. Convergence was lost, the visual axes of both eyes remaining parallel when the finger was made to approach the eyes in the median line; the internal rectus muscle of each eye, however, functioned perfectly in the associated motion toward right and left and also in the unassociated movements inward. In contradistinction to this, however, there were no upward movements in either eye when each was tested singly. The pupils were 3 mm. and were prompt to light but somewhat sluggish to accommodation stimuli. The fundi were normal, the nerves being of good tint. Corrected vision equalled 5—5 in each eye. The visual fields were normal, save for a slight concentric contraction for form and color in each eye.

"When questioned further regarding the nature of the vertigo of which he particularly complained, it was elicited that he had never lost consciousness in the attacks, but that he is unable to stand, on account of extreme giddiness. He frequently has uncontrolable laughing fits at the time. There are no spasms during the attack, and though he falls he is able to rise again at once."

This patient was referred to me for examination by Dr. Posey, Sept. 23, 1903. My notes made at that time are as follows:

There is no contraction of the visual fields tested by the hand. The irides respond promptly to light, but slowly in accommodation. The speech is indistinct and bulbar in character, and he has had difficulty in swallowing during the past three months. The saliva dribbles from his mouth constantly. The tongue is tremulous and possibly a little atrophied on the right side. The fifth and seventh nerves are not involved. Lateral movements of the eyeballs are good. He seems to have some slight impairment of downward movement of the eyeballs, as well as complete paralysis of upward movement. Attempts to look either upward or downward cause dizziness. Sensations for touch and pain are normal in all parts of the body. He complains of bad vision. The breath is very offensive and indicative of mercury. Voluntary power is good in all the limbs. He sways slightly when standing erect with eyes open, and still more so when eyes are closed. The biceps and triceps tendon reflexes and wrist reflexes are probably a little prompter than normal. The patellar reflex is much exaggerated on each side, but the Achilles reflex is normal on each side, and there is no ankle clonus. There is no Babinski reflex, the toes on the left side are not distinctly moved while those on the right side are flexed. There are no urinary disturbances.

Dec. 19, 1903. He has been having severe occipital headache during the past week. The saliva still dribbles from the mouth. When rising from a chair he lost his balance and fell to the floor. He has fallen before this. He does not show hemiasynergy. He says he can hear perfectly.

I received the following note from Dr. W. Zentmayer concerning the patient: "An examination Dec. 28, 1904, showed that the position of rest of both eyes was about 20° below the horizontal plane. All attempted conjugate movements except to the right were lost, and that to the right was limited. When not commanded he at times moves the eyes slightly to the left. The right palpebral fissure is decidedly wider than the left. The pupils are normal. Convergence power is lost."

Through the courtesy of Dr. M. H. Fussell, Dr. Zentmayer, and Dr. P. N. Moylon, I had an opportunity to examine this patient again on Jan. 22, 1905, at which time I made the following notes:

He has no headache and no dizzy spells, but his statements are not reliable. He has not had nausea or vomiting, at least since Oct. 1, 1904. Mentality is affected. Understands what is said to him, but in replying repeats his words several times. His speech is indistinct and he mumbles much in talking, and has not much modulation of his voice. Frequently he utters a peculiar bellowing noise and has forced movements of face, although it is hard to say whether they are more like laughing or crying. He is said to choke frequently in swallowing solids. Raises forehead well on each side and closes eyelids well, and draws up the lower part of each side of his face well. Masseter contracts well on each side, and there is no deviation of lower jaw on opening the mouth. Sensations for touch and pain are normal in the face. The tongue is protuded straight, is not atrophied, and shows no fibrillary tremors, and the movements are free. With finger in right ear he hears the voice when it is not very loud, and same is true when the right ear is tested; he is not deaf.

He is entirely unable to raise the eyeballs, but has slight conjugate lateral movement to the right and left, downward movements are much impaired. Hemianopsia is not present and the vision in upper and lower fields seems to be good.

If left eyelids are closed the right eyeball goes much further to the right, or if the right eyelids are closed the left eyeball goes to the right better than do the eyeballs when he tries to look to the right with both eyes at the same time. Movement of either eyeball separately, when one eye is closed, to the left is also much greater than when he tries to look to the left with both eyeballs at the same time. The associated lateral movements are therefore much more impaired than are the lateral movements of each eyeball separately. Ptosis is not present. Pupils are equal, and irides react promptly to light. Accommodation is not so distinct. Convergence is entirely lost. Movements of head are free. There is some tendency for the eyes to be directed downwards. Grasp of each hand is good. Movements of the upper extremity are good. Biceps and triceps tendon reflexes are exaggerated a little on each side. Sensations for pain and touch are normal in the upper limbs. Finger to nose test shows a little ataxia on each side. Sense of position is normal in each hand. Stereognostic perception is normal in each hand. Resistance to passive movements in the lowers limbs is normal. Voluntary power in the lower limbs is normal at all parts.

Patellar reflex is a little exaggerated on each side. Ankle clonus is not obtained on either side. Babinski's reflex is uncertain because the patient holds the foot very firmly. Achilles jerk is not obtained, also because he holds the foot firmly. He rises from the bed with some difficulty to stand on the floor, and sometimes mades two or three attempts before he can get on his feet. Is able to stand with feet together and eyes closed with very little sway. In walking he does not stagger except occasionally. His gait is very fair, either with eyes open or closed. Frequently in sitting down he looses his balance and falls backwards.

Summary: F. S., forty-eight years of age, (Sept. 22, 1904) had paralysis of upward associated movement with loss of convergence. Downward movement was possible, but was difficult. Lateral associated movements at this time were preserved. The man had been very alcoholic. The speech was bulbar in character, and swallowing had become difficult. The saliva dribbled from the mouth. Ataxia of station was present. Headache was severe at one period. By Dec. 28, 1904, associated lateral and downward movements had become much affected. On Jan. 22, 1905, I examined the patient again and found his mentality poor, speech more indistinct, deglutition difficult, associated upward movements lost, and all other associated ocular movements much impaired. The associated lateral movements were more impaired than the lateral movements of each eyeball separately. The upper limbs were slightly ataxic. Gait was fair, but frequently when he sat down he would lose his balance and fall backwards.

In this case paralysis of upward associated movement developed first, and was followed by paralysis of downward and of lateral associated movements.

Case 5. The report of this case was read by Dr. Zentmayer before the Ophthalmological Section of the College of Physicians.

A. S., male, aged fifty-seven years, butcher, was referred to me by Dr. Zentmayer, Dec. 3, 1904, with the following notes:

"He has had six children, all but two of these died before they were three years of age, one child is still living, aged twenty-nine years, the other died of phthisis. His first wife died of phthisis.

"O. D. 6/ix pt. O. S. 6/xii pt.

"One year ago he had lumbar pain and headache for two days. On the evening of Nov. 29, 1904, after returning from the theater, he found that he was dizzy and that his sight was blurred. He slept well, and the following morning had diplopia (?) on reading, also some nausea and frontal headache. He smokes and drinks to excess. His weight is 200 lbs., and he has not lost nor gained.

"He sits with his head thrown back. The eyes at rest drop slightly below the horizontal plane. There is apparently retraction of the upper lid. The upper lid touches the upper margin of cornea in O. D. In O. S., 1 mm. of sclera shows. On attempting to fix horizontal nystagmus and tremor of inferior fibers of orbicularis develop. Convergence is weak and nystagmic. Lateral conjugate movements are full but accompanied by short nystagmic jerks. Both conjugate and individual upward movement of globes are abolished. At times there is apparently some very slight upward movement of O. D. Right fissure 10 mm. Left 11 mm. Irides react to light and attempted convergence. O. S. iris somewhat sluggish to light. Ophthalmoscopic: peripheral lenticular opacities. Disc red gray with incipient neuritis (?). Veins dark and somewhat engorged. Arteries are a trifle small. Fields, O. D. white contracted to 20°, red contracted to 12°. O. S. white 20° above, 40° laterally, red 10° above, 15° laterally, no scotoma."

Notes made by me Dec. 3, 1904, are as follows:

He has had headache more than a year, and this he describes as a "kind of heavy feeling" in front of head and not a sharp ache. He drinks a large amount of beer and some gin, and smokes much. He has not been getting weak. He has not had any pains in his limbs, except in winter time he has had some rheumatic pains in his hands. He has poor appetite and indigestion.

He has slight drooping of each upper lid, and does not raise his eyes above the horizontal in attempting to look upward, and has then fine lateral nystagmic movements. The movements to the right and left seem to be fully normal, but toward the left he has slight nystagmic movements. Downward movements are normal. The pupils are equal. The reaction of the iris to light and in accommodation is good in each eye, except that the reaction to accommodation in the right eye is not quite so good as in the left eye. He converges with both eyes, but better with the left, and when he converges the upper lids tremble. Hearing for watch is excellent in each ear. The masseter contracts well on each side. There is no involvement of the facial nerve on either side. Sensation for touch and pain in the face and hands and lower limbs is normal. The grasp of the right hand is excellent, the left hand is on a splint because of a recent injury. The biceps and triceps reflexes of each upper limb are not distinct. The patellar reflex is weak on each side, but this probably is because it is impossible to get him to relax his limbs, and it is increased by reinforcement. The gait and station are normal, with eyes open or closed.

On Dec. 29, 1904, Dr. Zentmayer sent me the following note:

"Ten days after you saw A. S. there was noted a marked improvement in supraduction, and 4 days later he complained for the first time of diplopia, and a careful study of the movements of the eyes showed that there remained only a paresis of the left superior rectus. Yesterday all of the muscles (intra- and extra-ocular) seemed to have regained their full power. There is still glycosuria.

Summary: A. S., fifty-seven years of age, (Dec. 3, 1904) suddenly became dizzy and complained of blurred sight. He had used alcohol and tobacco to excess. Convergence was weak and nystagmic. Associated lateral movements were not limited, but were accompanied by short nystagmic jerks. Associated upward movements were lost. Reaction to light was preserved. The disc was red gray with incipient neuritis.

I saw the patient Dec. 3, 1904. He had been having headache. He had slight drooping of the upper lids. He had no signs of implication of his nervous system other than those mentioned.

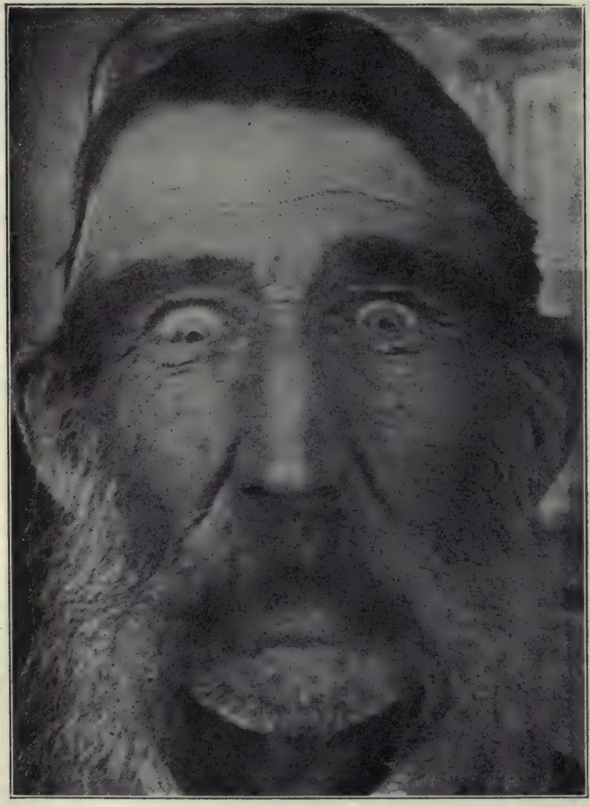

FIG. 11. Attempt to look upward. Forehead is wrinkled, but eyeballs do not move upward. (Case 6.)

Ten days later there was marked improvement in supraduction and four days later he complained for the first time of diplopia. There was then only paresis of the left superior rectus. By Dec. 28, 1904, all the ocular muscles had regained their full power.

Case 6. S. Hea., sixty-three years old, referred for examination by Dr. de Schweinitz, Dec. 14, 1904.

The patient is married and has six children. He attributes his condition to the atmosphere from oil works. He has

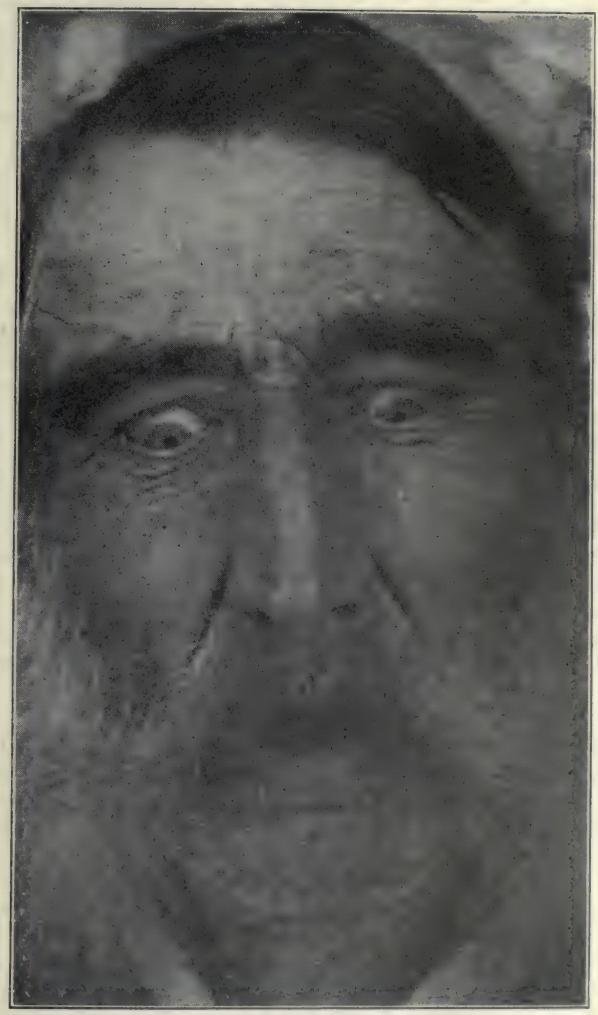

FIG. 12. Downward movement of eyeballs well performed. (Case 6.)

tremor, especially in the lower limbs. He denies all venereal diseases. He has been very alcoholic and has had difficulty in breathing. Has had headache every night about 8 o'clock, disappearing during sleep. Has bearing down pain across abdomen. Many members of his family have died from cancer. He weighed 181 lbs. a year ago, now weighs 145. No dizziness. Staggers in walking. Is weak in his whole body, "can't carry ten pounds."

He cannot look upward at all, but moves his eyeballs freely in all other directions. There may be some weakness of the right external rectus. The pupils are about equal, reaction to light is present in each eye, distinct but not very prompt. He can converge with each eye. Reaction of each iris in convergence and accommodation prompt and better than to light. No involvement of either facial nerve. Masseters contract firmly. Sensations for touch and pain seem to be normal everywhere. Can hear a watch with right ear only when the watch is pressed against the ear, cannot hear it with the left ear at all. Hears the voice, but says his hearing has become impaired. Soft palate moves freely, tongue normal. Grasp of each hand fair, but not powerful. Biceps and triceps jerks not prompt on either side. Patellar reflex is much diminished on each side. Ankle clonus is persistent on each side, more so on right. Babinski sign is uncertain, but probably present on each side. Sensations for touch and pain normal in the feet. He does not stagger when walking in the room or when standing, with eyes closed or open, but says he staggers on the street. Resistance to passive movements good in all the limbs. The tremor in the right lower limb seen while under my examination appears to be the result of ankle clonus.

Summary: S. H., sixty-three years old (Dec. 14, 1904), had been very alcoholic. He had tremor of the lower limbs, headache, slight ataxia, some subjective weakness of limbs, and complete paralysis of upward associated movements; all other associated movements were normal. Convergence was preserved. The patellar reflexes were diminished. Ankle clonus was present on each side, but Babinski's sign was uncertain.

Case 7. T. Roberts, male, a patient of Dr. de Schweinitz, was examined by Dr. C. K. Mills and myself about Nov. 12, 1904. The man was not ataxic. He could not see light, and had paralysis of upward and downward movements. Dr. de Schweinitz's notes, Nov. 29, 1904, are as follows:

"There is complete blindness, the ocular examinations showing choked disc, beginning to subside with the development of atrophy. He stated that severe headaches had been present before the blindness came on. There was also at times nausea. Headaches have been better, and I believe have been practically absent since the blindness, which he maintains appeared suddenly some months ago. So far as we can determine, there is entire loss of the upward and downward movements of the eyes, therefore failure of associated parallel movement upward and associated parallel movement downward. There is no failure in the power of the eyes to move in associated parallel direction directly to the right, or in a similar direction directly to the left, that is to say, no failure in dextroversion or levoversion. Movement of convergence is present, that is to say, if he is told to look at his own finger, which he rapidly approaches to within a few inches of his eyes, there is convergence of the eyes to this point."

Summary: T. Roberts had paralysis of associated upward and downward movements and was completely blind and had choked disc. He had had severe headache. Lateral associated movements were normal, and convergence was preserved.

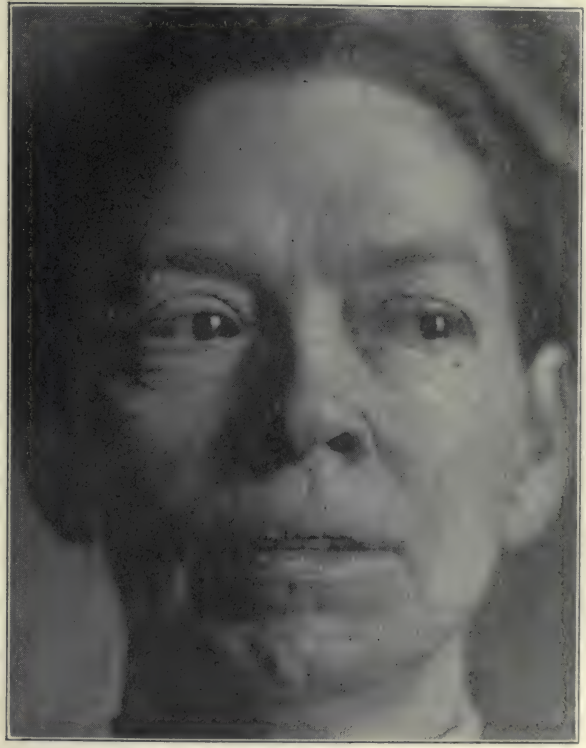

FIG. 13. Looking to the left. Slight weakness of left external rectus. (Case 8.)

Case 8. Mrs. McM., a patient of Dr. Wm. J. McConnell, aged fifty years, was seen by me in consultation with Dr. McConnell Jan. 15, 1905, when I made the following notes. The history and photograph were obtained from Dr. McConnell:

She has had 9 children, no miscarriages. Lungs and heart are normal. Urine has shown albumin and hyaline casts. In Oct., 1904, had her first convulsion, nature of which is not known. Had later another which she says began with numbness in left hand. Since first convulsion has had severe persistent occipital headache, occasionally nausea and vomiting, and she says dizziness. Some ataxia of gait has been observed. Complains that her sight is not so good as formerly. Last Monday (Jan. 9) had a third convulsion beginning, she said, with numbness of left hand, and since then mental state has been a little dull. She has lost flesh.

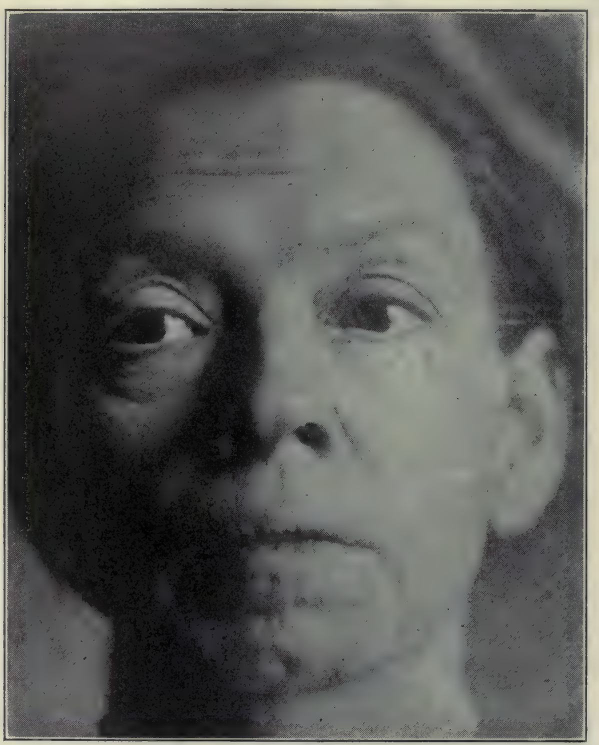

FIG. 14. Attempt to look upward. Wrinkling of forehead in attempt to look upward, but eyeballs do not move upward at all. (Case 8.)

Present condition. Partial ptosis which the patient cannot overcome. Each upper lid droops as low as the center of the pupil when she is looking directly forward. Paresis distinct in upward movement, raises the eyeballs only slightly above the horizontal. Some weakness apparently of left external rectus. Convergence completely lost. No paralysis of associated lateral movements. Movements to right and downward normal, and usually movements to left normal, but there may be some weakness of left external rectus, and occasionally when left eyeball does not go as far as it should to the left the right eyeball also does not go as far as it should to the left. Slight indication of nystagmus in looking to right. Pupils equal and reaction to light prompt. Movements of 7th and 5th nerve supply normal. Tongue protruded straight and not affected. No deafness to the voice. Sensation for touch and pain normal everywhere.

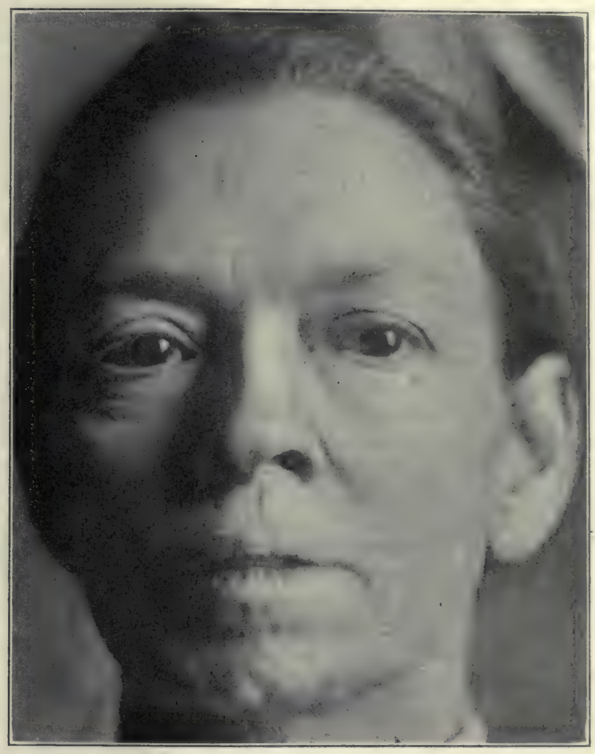

FIG. 15. Attempt to converge, showing power of convergence lost. (Case 8.)

Upper limbs not distinctly ataxic. Biceps and triceps tendon reflexes are not distinct. Movements of the upper limbs are normal. No loss of stereognostic perception nor of sense of position.

Gait slightly ataxic, sometimes falls toward left, sometimes toward right. Patellar reflex not exaggerated. No ankle clonus, Achilles jerks not distinct. No Babinski, toes seemed to be flexed. Voluntary power in all limbs fair, no paresis. No one has ever seen the patient in a convulsion.

I saw this patient again Feb. 1, 1905. The ocular palsies had disappeared, she could raise her eyeballs and upper lids fully, her intelligence was good, she walked well. She had been taking large doses of mercury and iodide. The case appeared to be one of syphilis.

Later the paralysis of associated upward movements returned but disappeared under the same treatment.

Summary: Mrs. McM., fifty years of age (Jan. 15, 1905), had convulsions, headache, nausea, vomiting, dizziness, ataxic gait, failure of sight. I found, Jan. 15, 1905, partial, bilateral ptosis, marked impairment of upward associated movements and loss of convergence; other asociated movements were preserved. The left external rectus was weak. Gait was slightly ataxic.

When I saw this patient again, Feb. 1, 1905, the ocular palsies had disappeared, and she could raise her eyeballs and upper lids fully. She had been taking large doses of mercury and iodide. Later the paralysis of associated upward movement returned and again disappeared under the administration of mercury and iodide.

Noteworthy in this case was the association of partial bilateral ptosis and paresis of the left external rectus with paralysis of upward associated movements; also the disappearance of the ocular palsies twice under the administration of mercury and iodide.

Case 9. W. B. was a patient of Dr. Wharton Sinkler, from whom the following notes were obtained. An opportunity was given to me to examine this patient.

W. B., insurance agent, aged twenty-eight years, was admitted Oct. 10, 1903.

Family history. His mother died from cancer. His father is living. Two brothers and three sisters are living and well.

Previous history. He had measles, pertussis, chicken pox, scarlet fever, diphtheria, and in 1900 syphilis.

He had not used alcohol or tobacco for 5 years. Before that time used both to some extent.

He has worried a great deal about business and felt that he was becoming nervous. May 10, 1903, he developed a right hemiplegia, affecting the arm first, then the leg, although the leg was never completely paralyzed, and he could walk around. Speech was not affected. Oct. 8 he developed diplopia.

On admission he complains of intense headache and diplopia, also of weakness in the right arm and leg, although he can walk and get around fairly well. He also complains of biliousness.

The bowels are regular, appetite has been very poor for the past few weeks, before that time it was very good. Sleep is good.

Heart and lungs are negative.

He has no gastric crises; no shooting pains or girdle sensation.

Reflexes are greatly exaggerated on the right side; slightly so on the left. Dynamometer, R=10. L=65.

The clavicles are very prominent and there is an enlargement of the inner end of the right clavicle. Interspaces are prominent.

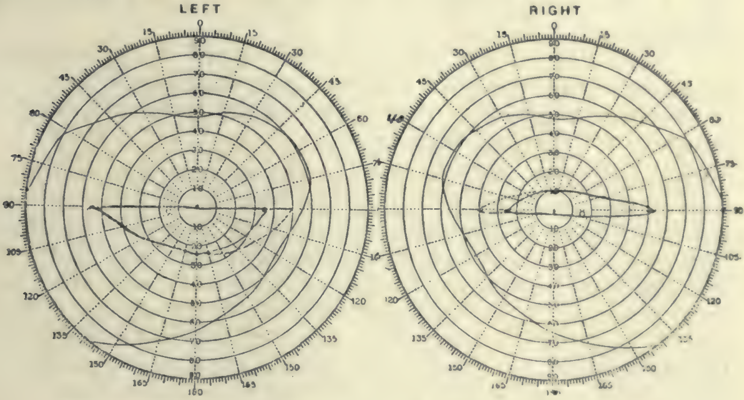

Fig. 16. Upward movement was abolished in the left eye, and was very slight in the right eye. Downward movement was lost in the right eye and was much impaired in the left eye, although it was greater than the upward movement of the right eye. Associated movements were nearly normal in each lateral direction. Convergence movements were lost. Rotation, fields of fixation. Right out = 50, right up = 10, right in = 25-40, right down = 0. Left out = 55, left in = 25-40, left down = 25.

Eyes. Report from Dr. De Schweinitz's clinic book:

O. D. 6/6+40.50, p.p 25; O. S. 6/6+4 O. S. 0.50 p.p 25. Paralysis of accommodation. Lateral outward rotations normal in both eyes; slight loss of inward rotation of each eye. Complete loss of downward movement of right eye, and partial of left eye. Nystagmic movements in both when rotated to extremes of fixation. No choked disc, although slight veiling of right disc; vessels normal. Right pupil—4 mm., faint reaction to light. Left pupil—2 mm., prompt light reaction. No convergence movement, but associated movements are nearly normal in lateral directions.

Nov. 7, 1903. Much improved. Almost all movements restored except downward of right eye, which is still deficient.

Treatment: massage, protiodide gr. 1-6 t.i.d., KI. gr. v increasing.

Oct. 19, 1903. For the past few days the patient has exhibited some mental symptoms, consisting of delusions. He thinks he has recently been married and has a child. That he has a contract to build a depot. That there is a jeweler's shop in the hospital, and that the goods are being auctioned off. At present there is decided mental deterioration.

Nov. 4, 1903. The patient to-day became sick at the stomach and vomited. He also had diarrhea and lost control of the bowels. He is still very much confused, and several times has appeared in the hall without any definite cause. He wanders about. His mental states is unimproved. KI and mercury are discontinued.

Nov. 6, 1903, The patient is discharged to-day somewhat improved. The eyes have recovered considerably from the loss of movements. Speech at present is quite thick and drawling in character. Mental state is poor.

Summary: W. B., aged twenty-eight years, had contracted syphilis in 1900. Right hemiplegia developed May 20, 1903, and in October of the same year he had diplopia. Headache became intense. Upward movement was abolished in the left eye, and was very slight in the right eye. Downward movement was lost in the right eye and was much impaired in the left eye, although it was greater than the upward movement of the right eye. Associated movements were nearly normal in each lateral direction. Convergence movements were lost. The pupils were unequal, and reaction to light in the right eye was faint, but prompt in the left eye. Optic neuritis was not present, but there was a little veiling of the right disc. Each ciliary muscle was completely paralyzed. Convergence was lost.

By Nov. 7, 1903, the patient was much improved, and almost all the ocular movements were restored, except the downward movement of the right eye.

ABSTRACTS OF THE CASES OF PARALYSIS OF UPWARD OR DOWNWARD ASSOCIATED OCULAR MOVEMENTS REPORTED IN THE LITERATURE.

The original papers, with three exceptions, were obtained and the abstracts were made by me from these.

Henoch[20]: The eyes were turned downward, and the child could not raise the eyes upward. Lateral movements were normal. Pupils were somewhat dilated and reaction was slow. Later were vomiting, drowsiness, paresis of the right arm, irregular pulse; still later, complete paralysis of the right limbs and right side of the face, deviation of the right eye inward. The lesions were tuberculous meningitis and tubercle of left posterior part of the corpora quadrigemina. It is not stated whether the third nerve nuclei were normal.

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[20] Henoch. Berliner klin. Wochenschrift, March 21, 1864, p. 125.

Wernicke[21]: After an apoplexy the symptoms were left hemiplegia, great limitation of upward and downward movements of the eyes, with normal lateral movements. There were no ptosis and no changes in the fundus. An apoplectic focus was found in the right optic thalamus, lenticular nucleus and anterior part of the right side of the corpora quadrigemina. The oculomotor nucleus was involved on the right side.

Priestley Smith[22]: Case 4. The symptoms were headache, vomiting, vertigo, diplopia. The eyes maintained a constant convergence toward a point two feet distant from the face, and the patient was unable to diminish to the smallest extent the convergence, nor could he increase it very much. Vertical movement upward above the horizontal plane was totally lost. Both eyes followed an object to the normal extent to the right or left. The patient steadily improved and he became able to look upward, but the ocular symptoms returned and the parallel conjugate movements were less free than at the former examinations. Tested by the horizontal and vertical movements of an object before the face, each eye was found to have the following range:

Outward, only a little beyond the median line, and much unsteadiness and jerking attended the effort.

Inward, to the full extent, but with a little jerking.

Downward, apparently to the normal extent.

Upward, no movement was possible above the horizontal line, and much jerking attended the effort. There was no necropsy.

Case 5. The symptoms were headache and vertigo, followed by sudden unconsciousness lasting one day, and supposed to be hysterical. One or the other eye deviated inward. The outward, inward and downward movements of the eyes were nearly or entirely to the full extent, but upward movements were not more than half the normal extent. The ocular symptoms later disappeared.

Nieden[23]: The symptoms developed suddenly. The patient, a man aged 45 years, became blind while bending over. Examination showed that vision was much impaired. The eyeballs could be moved promptly downward, outward or inward, but not upward beyond the horizontal line. When the attempt was made to look upward the eyeballs were drawn backward and forward in a peculiar manner. There were no fundus changes. The lesion was supposed to be a cerebral hemorrhage. Recovery in this case was complete.

Gowers[24]: The symptoms were headache, vomiting, weakness of the legs, a few convulsions. The patient's manner was suggestive of hysteria. She had well-marked double optic neuritis. The eyes moved freely in all directions but upward. When she tried to look upward the eyes moved very little or not at all above the horizontal line. The pupils were equal, 4 mm. in diameter, acting very little to light and not at all on attempts at accommodation. The patellar reflexes were much exaggerated. A very small tumor was found in the middle line behind the posterior quadrigeminal bodies, damaging these slightly, the velum and the adjacent part of the inferior vermiform process of the cerebellum.

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[21] Wernicke. Berliner klin. Wochenschrift, July 3, 1876, p. 394. "Lehrbuch der Gehirnkrankheiten." Vol. 2, p. 84.

[22] Priestley Smith. Ophthalmic Hospital Reports, 1876, Vol. 9, p. 22.

[23] Nieden. Centralblatt für prak. Augenheilkunde, July, 1880, p. 209.

[24] Gowers. Transactions of the Ophthalmological Society of the United Kingdom, Vol. 1, 1880-81, p. 117. A Manual of the Diseases of the Nervous System, Sec. Edition, p. 185.

Parinaud[25]: Case 4. A man, 67 years of age, had had attacks of polyuria and heaviness of the head 18 months. In one of these he had difficulty in standing and the vision was disturbed; he had a tendency to fall to the left. At the same time he had complete paralysis of upward and downward movement of the two eyes, except that the right eye could be moved upward a trifle. Movements to the left and right were normal. Convergence was lost. The upper lids were normal. The pupils were moderately contracted and a little unequal. The reaction to light was lost. The visual fields and central vision were normal. The pulse was slow. No necropsy was obtained.

Case 5. A woman, 20 years of age, had had headache, especially on the left side, about a month. Diplopia suddenly developed. A month later she was found to have complete paralysis of convergence. Lateral associated movements were normal, as were also downward movements, while upward associated movements were almost lost. Crossed diplopia was present. There was no lesion of the fundus, and no amblyopia. Several times numbness of the left lower limb occurred, and was followed by some weakness.

Ormerod[26]: A man, aged 44 years, had paresis of the upward movement of the eyes, most marked in the right eye, especially when the patient looked to the right. There was vertical nystagmus when he tried to look upward. The other ocular movements were normal. There was no definite neuritis. The symptoms had been present ten months, but he had had a similar condition fifteen years previously.

Otto Hope[27]: The symptoms were: Headache, general tremor occurring in attacks, right-sided deafness and vomiting. The patient was unable to raise his left eye above the horizontal, and the right eye he could raise only slightly; the downward movement of the eyes was much impaired. Lateral movements were normal. He had reflex rigidity of the pupil, and slightly choked discs. The limbs were not paralyzed. A tumor the size of a pigeon's egg was found in the corpora quadrigemina; the aqueduct was preserved only in the posterior half. The tumor was a telangiectatic sarcoma.

Reinhold[28]: The symptoms were vertigo, headache, paresis of each external rectus, vomiting, disturbance of vision without optic neuritis, bilateral ptosis and limitation of upward movement of the eyes, with nystagmus. A gliosarcoma of the pineal gland was found. The corpora quadrigemina and the aqueduct of Sylvius were compressed, but the tumor was easily separated from the former.

R. Thomsen[29]: The symptoms were failure of memory and intelligence, headache, vertigo, weakness, tremor, rigidity of the limbs, small and unequal pupils, iritic reflex to light lost on the right side and weak on the left, pallor of discs. Both eyes could be moved to the right, left or downward, but with nystagmus. The eyes could scarcely be raised above the horizontal, and the right was more affected. A gumma was found at the exit of the oculomotor nerves and growing into the peduncles and implicating the nucleus ruber, and especially the fibers of the right oculomotor nerve within the peduncle. Ependymitis diminished the size of the aqueduct of Sylvius considerably, but the gray matter about the aqueduct and the oculomotor nuclei appeared to be normal. Nissl's method was not employed. There seems to have been no compression of tissue by the gumma.

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[25] Parinaud. Archives de Neurologie, Vol. 5, No. 14, 1883, p. 145.

[26] Ormerod. Brit. Med. Journ., March 22, 1884, p. 564.

[27] Otto Hope. "Ueber einen Fall von Tumor der Vierhügel," Inaug. Dissertation, Halle a S., 1888.

[28] Reinhold. Deutsches Archiv. für klin. Med., Vol. 39, 1886, p. 1.

[29] Thomsen. Archiv. für Psychiatrie, Vol. 18, 1887, p. 616.

Nothnagel[30]: A male, 24 years old, presented the following symptoms: Headache, diplopia, paresthesia of left limbs, epileptic attacks, choked discs, paresis of the left upper and lower limbs, ataxia of the left upper limb, and objective disturbance of sensation in the left upper and lower limbs and left side of face. The left abducens was paretic. Upward and downward movements of each eye were much impaired. Lateral movements, except outward movement of the left eye, seem to have been good The left facial nerve was paretic for voluntary movements. There was no necropsy at the time of the report.

Samuel Gee[31]: The patient was six years of age. The symptoms were loss of appetite, vomiting, headache, ataxia, convulsions followed by inward squint of the eyes lasting three days, then the eyes began gradually to turn upward. About the same time the power of the limbs was lost. The head was retracted. Both eyes were turned upward, so that, the corneæ were concealed by the upper lids. The right eye also turned a little outward. There was constant vertical nystagmus, no ptosis. The discs were white, their margins blurred and their vessels small; *i.e.*, they were atrophic and probably secondarily to optic neuritis. Palsy of the limbs was very incomplete, and there was no rigidity.

A pulpy, gelatinous mass was found over the surface of the cerebellum above and behind the medulla oblongata, a large, bleb-like sac distended with fluid bulged out between the crura cerebri. The corpora quadrigemina were flattened and "distended over a mass of gelatinous pink new growth which formed the anterior projecting extremity of a mass of similar new growth filling the whole cerebellum." The growth was considered to be sarcomatous.

Evidently the eyes could not be directed downward in this case, because it is stated that it was difficult to get a view of the fundus on account of the position of the eyes.

Eisenlohr[32]: The symptoms were drowsiness, vertigo, later tremor of the left arm, dilatation of the right pupil, slow reaction of the pupils, no changes in the fundus; still later, limitation of movements of eyes, especially upward and downward, polyuria, headache, vomiting, stupor, ataxic gait; still later, choked discs, right-sided ptosis. A bullet wound was found in the right side of the corpora quadrigemina. A portion of the third nucleus was injured.

Lichtheim[33]: Case 1. In addition to the signs of brain tumor, one abducens became paralyzed, and was followed soon by paralysis of upward movement of both eyeballs, optic atrophy and rigidity of the pupils. A glioma occupied the whole of the corpora quadrigemina and had grown into the right optic thalamus.

Case 2. The symptoms were vertigo, ringing in the ears, paralysis of one abducens and paralysis of upward movement of both eyeballs. Choked discs were present, and later ataxia. The tumor was in the corpora quadrigemina and optic thalamus.

Case 3. The symptoms were headache, diarrhea, vomiting, ataxia and weakness of the right side of the body. The left abducens was paralyzed, and there was paralysis of upward movement of the eyeballs and optic neuritis. A caseous tumor was found in the corpora quadrigemina, not involving the anterior part of this structure.

The abducens paralysis was attributed to pressure upon the nerve at the base of the brain, and was present in all three cases.

Parinaud[34]: The patient, a man, became paralyzed in the left side of the face and in the left upper limb. The lower limb seems to have escaped, and the weakness of the left upper limb was of short duration, except in the fingers. He was almost completely unable to look upward, and lateral movement to either side was very imperfect. He was able to look downward without difficulty. Convergence was normal. The pupils were of normal size, and the iritic reaction was normal to light and convergence. There was no lesion of the fundus. He had partial left homonymous hemianopsia. Parinaud localized the lesion in the cortical center of the face and adjoining region.

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[30] Nothnagel. Wiener med. Blätter, 1889, No. 9, p. 131.

[31] Gee. Saint Bartholomew's Hospital Reports, Vol. 26, 1890, p. 106.

[32] Eisenlohr. Münch. med. Wochenschrift, May 20, 1890, p. 364.

[33] Lichtheim. Deutsche med. Wochenschrift, Nov. 17, 1892, p. 1043

[34] Parinaud. Annales d'Oculistique, 1892, Vol. 107, p. 283.

It is noteworthy that in this case reported by Parinaud himself convergence was normal.

Verrey[35]: The paralysis of associated upward and downward movement of the eyes developed suddenly, and was associated with a tendency to deviation to the left in walking. The symptoms lasted only a few weeks. A small hemorrhage was supposed to have occurred in the corpora quadrigemina or near this region.

Sharkey[36]: The patient, a man aged 43 years, became stuporous, deaf in the right ear, numb in the right half of the body, had right hemianopsia and paralysis of the superior rectus in each eye, and of the inferior oblique of the right eye. Both pupils reacted to light and accommodation, but sluggishly. There was no optic neuritis. Sensation was affected in the right side of the body. Speech was thick. Mr. Nettleship found that there was no movement of the eyes upward beyond the horizontal, and that downward movement was also defective, especially in the right eye. Concomitant lateral movements were full, but convergence was very defective, especially in the right eye. Irregular nystagmus was present in lateral and in upward movements. Fundi were normal.

Later, loss of power was noticed in the right side, and optic neuritis developed. A tumor was found in the corpora quadrigemina and upper part of the left cerebral peduncle.

Sauvineau[37]: A woman, aged 73 years, had an apoplectic attack with transitory right hemiplegia at the age of 70. Three years later she had another apoplectic attack with slight right hemi-paresis, and at the time of examination by Sauvineau she was a little weak in the right side. She did not have strabismus nor ptosis, and lateral movements of the eyeballs were normal, although they were associated with a little rotary nystagmus. Upward movement was lost in both eyes, and downward movement was feeble and associated with vertical nystagmus. Convergence was lost. At times she had diplopia. The pupils were equal and of normal size, and reaction in accommodation was normal. The light reflex was preserved, but feeble on the right side, and abolished on the left. No fundus changes were present, and the visual fields were normal. There was no hemianopsia. Sugar was present in the urine.

Schroeder[38]: The case was remarkable, as the paralysis of associated movement was downward. As cited by Teillais, a man 31 years old, was suddenly attacked with fever, headache, vomiting, convulsions of the limbs, complete ophthalmoplegia and amblyopia. After four or five weeks most of the symptoms disappeared, but there was complete paralysis of associated movement downward, and the eyes could not be moved downward below the horizontal. If he tried to look downward a spasm of the elevators occurred and the eyes were raised. No explanation was offered.

W. E. Bruner[39]: The chief symptoms were right hemiparesis and impaired vision. The movements of the eyes were perfectly normal in all directions except upward, in which direction there was scarcely any movement at all, not more than five degrees, but there was no diplopia. Slight nystagmus was observed when the patient attempted to look upward. No necropsy.

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[35] Verrey. Revue Médicale de la Suisse Romande., 1893, Vol. 13, p. 220.

[36] Sharkey. Brain, Vol. 17, 1894, p. 238.

[37] Sauvineau. Receuil d'Ophtalmologie, 1894, p. 592.

[38] Cited by Teillais. Annales d'Oculistique, Vol. 122, 1899, p. 19.

[39] Bruner. Columbus Med. Journ. Vol. 14, 1895, p. 505.

Hoesslin[40]: A boy, 9 years old, had had increased thirst and polyuria, headache, vomiting, ataxia, loss of muscular power in the lower limbs, and pupils dilated *ad maximum*. The eyes could be moved to the left, right and downward, but upward movement above the horizontal was impossible. He had bilateral choked discs. Both superior recti, both inferior oblique and the sphincter pupillæ were paralyzed. A spindle cell sarcoma was found growing from the pineal gland and corpora quadrigemina.

C. K. Mills[41]: The patient was unable to raise his eyes beyond the horizontal plane. The left eye diverged slightly, but there was no true lateral paralysis. The pupils were equal, reacting to light and in accommodation, both individually and consensually, but their movements were sluggish. The right field was concentrically limited to a decided degree, and slight concentric limitation was present on the left. The media and fundi were healthy. The central acuity of vision was 20-40 in the right eye and 20-30 in the left. Examination showed difficulty in speaking. The tongue was protruded slightly and a little to the left, and the patient showed some tendency to drooping. He complained of a general feeling of weakness in both legs. Both knee-jerks were exaggerated and a slight, probably spurious, ankle clonus was present on the left side.

Nothnagel[42]: Case 3, as given by Bach, there was bilateral ptosis. The movements of the eyes, especially of the left, were much impaired outwards and upwards. A tumor was found in the third ventricle, and the aqueduct of Sylvius was displaced. Pressure was probable upon the ocular nuclei and root fibers.

Basevi[42]: As given by Bach, associated paralyses were present (oblique superior and inferior rectus muscles, with disturbance of convergence). The corpora quadrigemina were scarcely recognizable.

Teillais[43]: There was complete paralysis of upward and downward movement of the eyeballs, with integrity of lateral movements, also complete paralysis of convergence, and yet the internal rectus of each side showed no impairment in lateral movements. The irides reacted to light, and the pupils were equal. There was no lesion of the fundus, no amblyopia, and the visual fields were normal. This condition lasted two months.

The man had syphilis and diabetes. The symptoms began in an apoplectic attack, and he remained almost unconscious a month without paralysis of the limbs. Only during the first day of his attack had he any difficulty in raising the eyelids. He had very few signs, other than the ocular, of involvement of his nervous system. He was very drowsy and slept much, and his intelligence was affected. He had dysarthria and the left upper limb was used in preference to the right, and the tendon reflexes on the right side were prompter. No necropsy was obtained in this case.

Poulard[44]: The patient had an apoplectic attack and loss of consciousness during six hours. Following this she had diplopia. Upward movement, and movements to right and left were normal, but associated downward movement was almost entirely lost; the pupils did not pass below the horizontal, and the patient was unable to follow an object downward. Convergence was much affected. There was some insufficiency of the left internal rectus, and also insufficiency in elevation of the left eye, although it was impossible to say whether the superior rectus or inferior oblique was at fault. In going down stairs the woman flexed the head *ad maximum*. The iritic reflex to light and visual acuity and visual fields were normal. Nervous symptoms, other than ocular, were slightly marked, the legs were a little stiff at times, and the tendon reflexes were exaggerated. There was no necropsy in the case.

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[40] Hoesslin. Münch med. Wochenschrift, March 31, 1896, p. 292.

[41] Mills. "The Nervous System and Its Diseases," 1898, p. 833.

[42] Bach. Zeitschrift für Augenheilkunde, Vol. 1, 1899, pp. 315, 455.

[43] Teillais. Annales d'Oculistique, Vol. 122, p. 19.

[44] Poulard. Revue Neurologique, 1901, p. 158.

Gordinier[45]: The patient was a man, aged 21 years. He had double optic neuritis, passing on to atrophy, intense and continuous headache, vomiting, dizziness, slow cerebration and gradual loss of memory, internal ophthalmoplegia, with double incomplete external ophthalmoplegia, marked cerebellar gait, coarse tremor of the hands, ataxia in the left leg, and choreiform movements. It is stated that there was slight convergence of eyeballs, and that the movements of the eyeballs were normal except upward and downward movements, both of which were practically abolished, and that there was slight double ptosis.

A neuroglioma, measuring 4x3x2½ cm., was found projecting from the superior worm of the cerebellum. It had involved by actual ingrowth the posterior part of the corpora quadrigemina on each side, and had destroyed almost completely the interior of the right posterior colliculus, leaving but a superficial shell of cortex; the left posterior colliculus being similarly, although much less, involved. The anterior colliculi did not appear to be affected. The aqueduct of Sylvius was occluded, the third nerve nuclei were distorted, and the left nucleus contained a less number of cells than its fellow of the opposite side. The nucleus of each side contained numerous atrophic cells devoid of processes and without nuclei; other cells had lost their processes and showed much alteration. The dorsal part of the oculomotor nuclei was most affected.

Crouzon, Marie, Babinski[46]: The patient, a man, had an apoplectic attack lasting seventeen hours, and became blind. During the attack he did not have stertorous breathing nor evacuation of the bladder or bowels. The coma disappeared suddenly, but the patient was delirious for several weeks, and did not recognize persons or objects. The speech was slow and hesitating and the visual fields were contracted. The eyeballs were turned upward; when he threw his head as far backward as possible and followed with his eyes a finger slowly lowered the eyeballs moved downward, but if his head were erect and he attempted to look at his feet he bent his head forward and the eyeballs went forcibly backward.

Raymond and Cestan[47]: Case 3. A man, 43 years of age, had left sensori-motor hemiplegia. Eight days after a severe mental shock his speech became affected and he became paralyzed on the left side and had diplopia. The tendon reflexes on the left side were exaggerated and Babinski's sign was obtained. There was no ptosis. The fundus was normal. The pupils were equal, and the reaction to light and in accommodation was normal. The man had paralysis of associated ocular movements both toward the left and toward the right, but more toward the left There was also marked paralysis of upward associated movement. Downward associated movement was almost normal. Convergence was slightly affected. No necropsy was obtained. This condition had existed ten years.

Noguès and Sirol[48]: A woman, 50 years of age, had been in bed about three weeks because she had lost her appetite, had difficulty in sleeping, had lassitude and was unable to work, had chills, and probably fever, with palpitation. She noticed the first day she arose from her bed that she could not look upward. She was found to be unable to look above a certain point a little below the horizontal. Downward and lateral movements and convergence were normal. When the upper lids were lowered she could raise her eyes above the point she could reach if her eyes were open. The fundus was normal. There seemed to be no other signs of organic disease of the nervous system. Because of the absence of an apoplectic seizure and of other symptoms of an organic nature, and of paralysis of convergence, and because of certain hysterical manifestations and of the probably sudden development of the paralysis, Noguès and Sirol regarded the paralysis of upward associated movement as hysterical. It seems to me the hysterical nature of this case has not been established.

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[45] Gordinier. JOURNAL OF NERVOUS AND MENTAL DISEASE, Vol. 28, 1901, p. 543.

[46] Revue Neurologique, 1901, p. 428.

[47] Raymond and Cestan. Revue Neurologique, 1901, p. 70.

[48] Noguès and Sirol. Revue Neurologique, 1901, p. 290.

Kornilow[49]: Case 1. A child, 6 years old, had complete paralysis in upward and downward movement, some impairment of convergence, and slight slowness in the reaction in accommodation and light. Movements of the eyeballs laterally were normal. Recovery in this case was almost complete.

Case 2. A child, 4 years old, had slight bilateral ptosis, and the left eyeball was turned somewhat inward. Movement of the eyeballs toward the right was almost normal, downward entirely normal, but toward the left the movement was much impaired, especially in the left eye, and upward associated movement was almost impossible. Convergence was normal.

The first case was supposed to be one of poliencephalitis superior of Wernicke; the second, one of tubercle.

In the first case there was slight weakness of the lower part of the right facial nerve supply and of the right hypoglossus; in the second, paresis of the right facial nerve and of the right upper and lower limbs. Ataxia was present in both cases.

Posey[50]: Case 1. Given in detail by me as Case 4 (Frank S.) of my report.

Case 2. The patient, a woman, was 65 years of age. She had had an attack of apoplexy and was hemiplegic on the right side. Neither eye could be rotated above the horizontal plane, though the other movements, including convergence, were preserved. The reaction of the irides to light and in accommodation was sluggish. The pupils were nearly equal, 3 mm. and 2½ mm., respectively. The fundi were normal. Corrected vision equaled 5-6 in each eye. There was no hemianopsia. The condition was unchanged after two years.

J. Porter Parkinson[51]: A girl, 11 years of age, began to stagger when 7 years of age. Sight failed. Headache did not occur. Movements of hands were incoördinate. The deep reflexes were increased. Hearing in the left ear was slightly impaired. Reaction of pupils was sluggish to light and in accommodation. The upward and lateral movements of the eyes were limited, especially the former. Discs were pale, but there was no evidence of optic neuritis. Speech was slow and hesitating. Necropsy was not obtained.

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[49] Kornilow. Deutsche Zeitschrift für Nervenheilkunde, Vol. 23, 1903 p. 417.

[50] Posey. Annals of Ophthalmology, July, 1904.

[51] Parkinson. The British Journal of Children's Diseases, Jan., 1905, p. 23.

Note: Two cases of paralysis of associated upward and downward movements, but not of associated lateral movements, have been reported by Gruner and Bertolotti too late to be included in this paper (*Nouvelle Iconographie de la Salpêtrière,* 1905, No. 2, p. 159). In one case a tumor of the tegmentum of the cerebral peduncles in the region of the corpora quadrigemina was supposed to be present; in the other a tubercle had destroyed the oculomotor nuclei. A clinical case by W. A. Turner (*Brain,* 1898, Vol. 21, p. 341) also should be included.

TABLE SHOWING CASES OF PARALYSIS OF ASSOCIATED UPWARD OR DOWNWARD MOVEMENT.

	Paralysis	Pupils Dilated	Fundus	Lateral Movements	Ptosis	Convergence	Result of Lesion.	Other Symptoms,	Third nuclei.
Henoch 1864	Upward movement	Reaction slow		Normal Later deviation of right eye inward Eyes turned downward			Tub. meningitis and tubercle of l. post. part of corp. quad.	Vomiting, drowsiness, paresis of r. arm, and later, of both r. limbs and r. side of face	Condition not stated.
Wernicke 1876	Upward and downward		No changes	Normal	None		Apoplectic focus in r. optic thal., len. nuc. and ant. part r. side corp. quad.	Apoplexy L. hemiplegia	Involved on r. side
Priestley Smith 1876 Case 4	Upward			Normal Later imperfect Downward normal			Result not stated	Headache, vomiting, vertigo, diplopia, constant convergence of eyes	
Priestley Smith 1876 Case 5	Upward			One or the other eye deviated inward Outward, inward and downward nearly or entirely normal			Ocular symptoms disappeared	Headache, vertigo, temporary unconsciousness	
Nieden 1880	Upward		No fundus changes	Downward, outward and inward normal			Recovery complete, lesion supposed to be hemorrhage	Attempts to look upward caused eyes to be drawn backward and forward Vision disturbed	
Gowers 1880-'81	Upward	Acted very little to light, and not at all to accom.	Double optic neuritis	All directions free but upward			Small tumor in middle line behind post. quad. bodies, damaging them slightly	Headache, vomiting, weakness of legs, convulsions, patellar reflexes exaggerated	

Parinaud 1883 Case 4	Upward and downward	Moderately contracted and a little unequal Reaction to light lost		Normal	None	Lost	No necropsy	Polyuria, heaviness of head, difficulty in standing, vision disturbed
Parinaud 1883 Case 5	Upward		No lesions	Normal, as also downward		Lost		Headache, diplopia, numbness of lower limbs, followed by weakness
Ormerod 1884	Upward		No definite neuritis	Other ocular movements normal				Nystagmus looking upward Had a similar attack 15 years previously
Hope 1888	Upward, downward much impaired	Rigidity of pupil	Slight choked discs	Lateral movements ments normal	Bilateral	Said by Bach to be preserved	Sarcoma in the corp. quad. aqueduct preserved only in post. part.	Headache, gen. tremor, r. deafness, vomiting
Reinhold 1886	Upward		No optic neuritis				Gliosarcoma of pineal gland Corp. quad and aque. compressed	Vertigo, headache, paresis of each ex. rectus, vomiting, disturbed vision, nystagmus attempting to look upward
Thomsen 1887	Upward	Small and unequal Reaction to light affected	Pallor of discs	Right, left and downward normal, but with nystagmus			Gumma involving intramedullary 3d nerves	Failure of memory and intelligence, headache, vertigo, weakness, tremor, rigidity of limbs

TABLE SHOWING CASES OF PARALYSIS OF ASSOCIATED UPWARD OR DOWNWARD MOVEMENT.

Nothnagel 1889	Upward and downward		Choked discs	Lateral movements except outward movement of the left eye seem to have been good				Headache, diplopia, paresthesia of left limbs, epileptic attacks, paresis of left limbs, ataxia of left upper limb	
Gee 1890	Downward		Discs white, margin blurred vessels small, atrophy following neuritis		None		Sarcoma involving corp. quad.	Vomiting, headache, ataxia, convulsions, weakness of limbs, retraction of head. Both eyes turned upward, vertical nystagmus	
Eisenlohr 1890	Upward and downward	R. pupil dilated Reaction slow	Choked discs		Right ptosis		Bullet in right side corp. quad.	Drowsiness, vertigo, tremor of l. arm, polyuria, headache, vomiting, stupor, ataxia	A portion 3d nuc. injured
Lichtheim 1892 Case 1	Upward	Rigidity of pupils	Optic atrophy	One 6th paralyzed			Glioma of corp. quad. extending into r. optic thal.	Signs of brain tumor	
Lichtheim Case 2	Upward		Choked discs	One 6th paralyzed			Tumor in corp. quad and optic thal.	Vertigo, ringing in ears, ataxia	
Lichtheim 1892 Case 3	Upward		Optic neuritis	Left 6 paralyzed			Caseous tumor in corp. quad.	Headache, diarrhea, vomiting. weakness of r. side of body	

Parinaud 1892	Upward	Normal size Reaction to light and con. normal	No lesion	Lateral movements imperfect. Downward good		Normal	No necropsy	Paralysis of left face and left upper limb (of short duration) Partial l. homonymous hemianopsia
Verrey 1893	Upward and downward						Recovery	Tendency to go to the left
Sharkey 1894	Upward, downward defective	Reaction to light preserved, and accom. but sluggish	Optic neuritis	Lat. movements good		Con-verg. defective	Tumor of corp. quad and cerebral ped.	Right deafness, numbness r. half body, r. hemianopsia, sensation r. half body disturbed, speech thick, nystagmus, loss of power r. side
Sauvineau 1894	Upward, downward feeble	Pupils equal, of normal size Reaction in accom. normal L. reflex affected	No fundus changes	Lat. normal, but nystagmus	No ptosis	Conv. lost	No necropsy	Apoplectic attack, transitory r. hemiparesis, diplopia
Schroeder 1894 Cited by Bach and Teillais	Downward							Fever, headache, vomiting, convulsions, at first complete ophthalmoplegia Attempt to look downward caused spasm of elevators

TABLE SHOWING CASES OF PARALYSIS OF ASSOCIATED UPWARD OR DOWNWARD MOVEMENT.

Bruner 1895	Upward			Lat. and downward normal				R. hemiparesis, impaired vision, slight nystagmus attempting to look upward
Hoesslin 1896	Upward	Pupils much dilated	Choked discs	Lat. and downward good			Sarcoma growing fr. pineal gland and corp. quad.	Increased thirst, polyuria, headache, vomiting, ataxia, loss of power in lower limbs
Mills 1898	Upward	Pupils equal, reacted to light and accom, but sluggishly	Fundus normal				No necropsy	Slight difficulty in speaking, tongue protruded to left, weakness of legs
Nothnagel Cited by Bach	Upward and outward				Bilat. ptosis		Tumor of 3d ven. aque. displaced. Pressure prob. on 3d neuclei	
Basevi Cited by Bach 1899	Downward					Con. affected	Lesion of corp. quad.	
Teillais 1899	Upward and downward	Reaction to light preserved	No fundus changes	Lat. normal	Ptosis 1 day	Con. lost	No necropsy	Apoplexy, unconsciousness, drowsiness, impaired intellect, dysarthria
Poulard 1901	Downward	Reaction to light normal		Upward and lat. normal L. int. rectus affected		Con. impaired	No necropsy	Apoplexy, diplopia

Gordinier 1901	Upward and downward	Internal ophthalmoplegia	Optic neuritis	Lat. normal	Double ptosis	Con. slight	Neuroglioma invading post. part. corp. quad. Aq. occluded	Headache, vomiting, dizziness, impaired mentality, cerebellar gait, tremor	Third nuclei distorted
Cruzon Babinski Marie Parinaud 1901	Downward							Apoplexy, delirium, slow speech	
Raymond and Cestan 1901	Upward and lat., both r. and l.	Pupils equal Reaction to light and accom. normal	Fundus normal	Downward almost normal	No ptosis	Con. little affected	No necropsy	Left sensori-motor hemiplegia, affected speech, diplopia, exag. tendon reflexes, Babinski	
Noguès Sirol 1901	Upward		Fundus normal	Downward and lat. normal		Con. normal		Insomnia, lassitude	
Kornilow 1903 Case 1	Upward and downward	Reaction to light and accom. slow		Lat normal		Con. impaired	Recovery	Weakness right 7 and 12, ataxia	
Kornilow 1903 Case 2	Upward and left lateral			Left eye turned inward, r. almost normal, down normal, but to left impaired	Bilat. ptosis	Con. normal		Paresis right 7, right upper and lower limbs, ataxia	
Posey Case 1 is given below by me as Case 4, Sherry									

TABLE SHOWING CASES OF PARALYSIS OF ASSOCIATED UPWARD OR DOWNWARD MOVEMENT.

Case 2 1904	Upward movement	Reaction slow	Fundus normal	Good	None	Preserved		Apoplexy and right hemiplegia	
Parkinson 1905	Upward	Slow to light and accommodation	Discs pale but no optic neuritis	Lat. movements limited				Ataxia, exaggerated reflexes, slight impairment of hearing in left ear, speech slow and hesitating	

ORIGINAL CASES

Case 1 Galik	Upward	Reaction good	Optic neuritis	Lat. movements impaired	None	Preserved	Tubercle on left side of pons	Right hemiplegia of limbs, left palsy of face, involvement of each ext. rectus and left palsy of masseter, exaggeration of right tendon reflexes, hearing affected on left side, left neuroparalytic keratitis	3d nuclei not involved
Case 2 Moore	Upward	Prompt in each eye		Nystagmus in lateral movements Paralysis of movement of both eyeballs to right and paresis of both to the left	Slight ptosis	Preserved but impaired	Glioma of right side of pons	Weakness of each 7, left paresis of tongue, right paralysis of muscles of mastication, paralysis of left limbs, bulbar speech	3d nucleus much degenerated
Case 3 F. D.	Upward downward and lateral	Reaction prompt to light		Inward deviation of right eye	None	Preserved tho' not normal	Hemorrhage in r. tegmentum of pons	Paralysis of r. side of face, of r. side of tongue and of r. muscles of mastication Paresis of left limbs	

TABLE SHOWING CASES OF PARALYSIS OF ASSOCIATED UPWARD OR DOWNWARD MOVEMENT.

Case 4 Sherry (Reported by Dr. Posey)	At first only upward, later also lateral movements and downward impaired	Reaction prompt to light	Normal, at least in early stage of disease	Later conjugate lateral movements much impaired	None	Lost	Living	Impaired mentality, ataxia of limbs, some exaggeration of tendon reflexes, tendency to fall backward when he sits down
Case 5 A. S. (Reported by Dr Zentmayer)	Upward	Right reaction to light good, left sluggish, also good in accom.	Incipient neuritis (?)	Good, but nystagmic	None	Weak	Recovery with paresis of only left superior rectus, later recovery of this muscle	Dizziness, headache, no ataxia, glycosuria
Case 6 S. H.	Upward	Reaction present not very prompt to light Pupils equal		No impairment	None	Preserved	Living	Impairment of hearing, some general weakness, ankle clonus on each side, especially on the right
Case 7 Roberts	Upward and downward		Choked discs	No impairment	No ptosis	Preserved		Headache
Case 8 Mrs. McM.	Upward	Reaction prompt		Left ext. rectus weak (?); no paralysis of associated lat. movement.	Bilateral	Lost	Recovery twice, with relapses	Convulsions, headache, nausea, vomiting, dizziness, ataxia
Case 9 W. B.	Upward and downward	Unequal, reaction feeble in right eye	Veiling of right disc	Nearly normal	None	Lost	Almost all the ocular movements were restored except the downward movement of right eye	Right hemiplegia, headache

PSEUDO-SCLEROSIS (DIFFUSE SCLEROSIS),

WITH THE REPORT OF A CASE WITH NECROPSY.*

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WITH PATHOLOGIC REPORT AND REMARKS.
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In 1883 Westphal[1] reported two cases presenting a symptom-complex resembling that of multiple sclerosis, but in which microscopic examination of the brain and cord gave negative results. To this condition was given the name pseudo-sclerosis. Since this time a number of cases (14) presenting this condition have been reported, mostly by German observers. It will be noted in reading the abstracts of these cases, which are given below, that the brain and cord in a number of them were unusually firm. A similar condition of hardness, although possibly more marked, is observed in another group of cases also presenting a symptom-complex resembling that of multiple sclerosis, in which microscopic examination of the brain and cord shows a proliferation of neuroglia and vascular disease. To this condition the term diffuse sclerosis has been applied, and a number of cases have been reported, mostly by German authors.

As will be shown further on, some authorities are coming to the opinion that the same pathologic condition is present in both pseudo-sclerosis and diffuse sclerosis, the former being merely a milder degree of the latter. To call attention to this possible relationship is one of the objects of this paper.

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* Read before the Philadelphia Neurological Society, April 25, 1905.

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[1] Archiv. für Psychiatrie, vol. xiv, 1883, p. 87.

The pathologic condition now known as diffuse sclerosis has been frequently noted in the past, especially in the brains of idiots, and cases occurring in adults were reported years ago, although not under this title; but it is only recently that attempts have been made to erect on this basis a distinct disease.

Probably the first case was reported in 1849 by Duplay[2] to the Biologic Society of Paris. The patient had suffered for a number of years with paralysis and contractures of the lower extremities, probably brought on by overexertion and sexual excess. After a while the bladder, rectum and tongue became paralyzed, the mental condition being reported as good. At the autopsy the internal membranes were found to be opaque, but not adherent to the cortex; the gray matter was normal, but two-thirds of the white matter was sclerosed, more so toward the ventricles. This condensation was equal in extent on both sides. The tissue was yellowish and cloudy. The optic chiasm and the striate bodies were also of greater consistency than normal. The cord was small and atrophic.

In 1854, under the title of "A Case of Cerebral Sclerosis," M. Hirsch[3] reported a case and stated that several others have reported cases of chronic induration of the brain tissue. His patient was a man, 53 years of age, who had been suffering for several years. At the autopsy the principal conditions found were an injection and infiltration of the pia-arachnoid, which was easily separated from the cortex, and a toughness and dirty-white color of the brain substance; there was very little cortex, the tough, dirty-white material taking its place; in some places the brain seemed cartilaginous. The ventricular lumen was small, the striate bodies and optic thalami were atrophic and tough. In the left frontal lobe several spots of sclerosis differing from the rest of the substance in luster were seen. The pons, medulla and peduncles were also indurated. The basilar arteries were sclerotic. The condition of the spinal cord was similar to that of the brain. The clinical symptoms were disturbances of sensation, headache,

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2. Schmidt's Jahrbuch, 1850, lxvii, p. 388.
3. Prag. Viertel Jahrsch. f. prakt. Heilk., 1854, 124.

dizziness, disturbances of motion as convulsive movements and contractures, paralysis of one hypoglossal nerve and of the bladder and anal sphincters, and causeless laughter and crying.

The first American case was probably reported by Echeverria[4] in 1869. His patient was a man aged 69 years, who had been an epileptic. His symptoms for several months before his death consisted of dizziness when he attempted to walk, weakness of the tongue, and paralysis of the lower limbs with contractures and involuntary muscular contractions. There was also pain in the legs. A bedsore and an eruption of herpes zoster developed. At times he was confused, at other times he would lose the power of articulating and swallowing, while at other times these processes and his mentality were intact. At the autopsy, in addition to aneurism of each vertebral artery, the cortex of the brain was found to be anemic and yellowish, and the brain substance was sclerosed, especially along the superior and inferior marginal convolutions, and both sides of the fissure of Sylvius. The medulla and the cord were also involved. Microscopically, the nerve elements were found to be diminished in number, their places being taken by proliferating connective tissue cells and fibers.

After this some other cases were reported, notably by Kelp,[5] and in 1878 A. Strümpell[6] wrote an exhaustive paper on the subject. He defined it as a condition in which the abnormal hardness and consistency of the brain, as a whole or in its greater part is noticeable on macroscopic examination. As causes he mentions hypertrophy of the brain, usually associated with idiocy; atrophy of the brain, senility, alcoholism, saturnism, and acute and chronic inflammatory diseases. He reports a case in an alcoholic and believes the sclerosis to have been due to a chronic diffuse interstitial encephalitis. He concludes that a definite disease can not be described from the reported cases.

After Echeverria, probably the next American cases were reported by Bullard.[7] In 1897 Heubner[8] reported a case and endeavored to set up a definite clinical picture. He describes it as a disease characterized by the appearance of paralysis and psychic disturbances. A spastic paresis of the legs first develops, which gradually extends to the arms; at the same time apathy and dementia appear and may terminate in idiocy. In the end a general paralysis develops; in some cases dysarthria and dysphagia occur. Optic neuritis was seen in one case. Etiologically he believes that hereditary syphilis and cerebral trauma play active parts.

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[4] "A case of Sclerosis of Both Third Anterior Frontal Convolutions Without Aphasia," Med. Record. 1869, iv, p. 1.

[5] Deutsches Arch. f. klin. Med., 1872, x, p. 224.

[6] Arch. f. Psych., 1878, ix, p. 268.

[7] "Cortical Sclerosis of the Brain in Children," W. N. Bullard, Jour. Nerv. and Ment. Dis., 1890, xv, 699.

[8] Charité Annalen, 1897.

In 1900 H. Weiss[9] reported a case and gave an exhaustive description of the disease, with full bibliography. His patient was a woman, 37 years of age, who had had symptoms of acute meningitis, associated with a suppurating middle ear. Two years after this attack she developed stiffness and weakness of the legs, painful spasms, hesitating speech, emaciation, increase of all the reflexes, increased mechanical irritability of the muscles. There was no involvement of the cranial nerves, sphincters or of sensation. He lays especial stress on the universal rigor which developed whenever she attempted to move. This he described as follows: When quiet, all the voluntary muscles are flaccid, with probably a slightly higher tonicity than normal; as soon, however, as movement is attempted the muscles become rigid, and the rigidity slowly increases to a maximum and then decreases in the same way. This rigor can be produced also by passive movements. All the muscles (trunk, extremities, larynx, eye, etc.) are affected. Every movement is laborious, hesitating and dragging. Speech is difficult because of spasm of the vocal cords. In addition to these symptoms there was apathy and gradually developing dementia. He considered the condition to be due to the previous meningitis. According to Weiss, other symptoms which may be present are vertigo, headache, ideas of grandeur as in paretic dementia, sudden changes of mood, the patient passing from a happy state into crying attacks; strabismus, inequality of the pupils, myosis, slow reaction of the iris to light, fibrillary tremors, general muscular atrophy, and tremor on movement.

In discussing the etiology, he states that the cases are to be divided into those existing before birth and those arising after birth. In the former class, the anatomic changes show that the child has had meningitis in utero associated with cerebritis, the nerve elements having been destroyed in large numbers. A meningo-encephalitis can also produce it in an adult; other factors are syphilis, either acquired or hereditary, individual predisposition, heredity, overexertion, traumatism, alcohol and epilepsy.

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[9] Obersteiner's Arbeiten, vol. vii, 1900, p. 245.

Patient.—The patient whose case is now reported was admitted to the Philadelphia General Hospital Oct. 28, 1900. The man was a negro, 57 years of age, and a laborer by occupation. His chief complaint was "tremor and weakness of the extremities."

History.—There was nothing to note in the family history. Until six months before admission, when his present illness began, he had been a healthy and hard-working man. Until four years previous to admission alcohol had been used to excess. Syphilis was denied. The autopsy, however, showed that probably he had had it. Gonorrhea he had had once. The first symptoms noticed were progressive loss of strength, staggering and a tendency to trip over small objects. He stated that at times he became giddy and had fallen. He had suffered from dyspnea on exertion for the past twenty years.

Examination.—The heart, lungs and other viscera were apparently normal. The urine was normal. There was ataxia of both arms, also a coarse tremor, which was intensified by voluntary movements, such as lifting a glass. The gait was slightly staggering and spastic. The knee-jerks were increased, ankle clonus was absent, but the Babinski reflex was present on the right side. The pupils were equal, did not react to light, but did in accommodation. There was no other involvement of any of the cranial nerves nor of the bladder or rectum. Sensation was normal. A diagnosis of multiple sclerosis was made.

March 1, 1901, the following notes were made: There is no scanning speech. Tremor of the tongue is not present when the tongue is protruded, but there is a marked tremor of the muscles of the upper part of the face at this time. When the arms are extended a coarse tremor is developed. Extension of the legs also causes a tremor to develop in them, which ceases when the patient attempts to touch anything with his foot. This movement, however, is ataxic and there is slight ataxia of both arms. Nystagmus is absent. The gait and station are fairly good. The knee-jerks are much exaggerated, and ankle clonus is present on the right side. The Babinski reflex can not be elicited.

July 5, 1902, it was noted that for several days previously the patient had been noisy and had refused to eat, asserting that the food had been poisoned. By July 19 these symptoms had disappeared. This was the only time that the patient's mental condition occasioned any comment, he having been up and about since admission. July 28 he was suddenly taken with a convulsion, which lasted an hour. He then became comatose and remained so until he died, on July 29.

Assuming that diffuse sclerosis and pseudo-sclerosis are distinct conditions, Frankl-Hochwart[10] has made an elaborate attempt to formulate points of distinction in their symptomatology. He compared 13 cases of so-called pseudo-sclerosis with 22 cases of diffuse sclerosis with the following results:

ETIOLOGY.

1. *Age.*—The majority of cases of both occur in early life; only 3 cases of diffuse sclerosis are recorded in which the symptoms appeared after 40.
2. *Heredity.*—A history of various forms of nervous disease in the family may be obtained in either condition. This, however, seems to be somewhat more frequent in pseudo-sclerosis than in diffuse sclerosis.
3. *Traumatism.*—No case of pseudo-sclerosis can be traced to this. Eight cases of diffuse sclerosis have such a history; 5 head injuries, 1 fall from a tree, 1 a lightning stroke, and 1 difficult labor.
4. *Lead Poisoning, Syphilis and Typhoid Fever.*—These are mentioned as causes of pseudo-sclerosis; alcoholism, meningitis, nephritis and syphilis among those of diffuse sclerosis.

SYMPTOMATOLOGY.

Paralytic Symptoms.—These are the same in both conditions, i. e., either paralysis or paresis, which may be transitory and change its location; it may be hemiplegic in type.

The Gait.—Either an ataxic or spastic gait is common to both affections, in some cases of diffuse sclerosis, however, walking is impossible on account of the spasms caused by the effort to walk.

Contractures.—These are common to both conditions and may be either transitory or permanent. The muscles of the back of the neck may be affected.

Tremor.—This is constantly present in pseudo-sclerosis and in two-thirds of the cases of diffuse sclerosis. The intention tremor is the more common; in pseudo-sclerosis this is in the nature of a to-and-fro movement, which does not occur in diffuse sclerosis. The head, face and tongue may also be the seat of tremor.

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[10] "Zur Kenntniss der Pseudo-Sclerose," Obersteiner's Arbeiten, 1903, x, p. 1. (A bibliography is appended in the monograph.)

Pain and Paresthesiæ.—These occur equally.

Mentality.—In pseudo-sclerosis mental disturbances are the rule but not as common as in diffuse sclerosis, not one of the patients with this latter condition having a normal mind. Epilepsy, headache, vertigo, causeless laughter and crying, syncope and apoplexy occur equally in the two affections. Paralysis of the tongue, either bilateral or unilateral, is noted more frequently in diffuse sclerosis.

Tendon Reflexes.—These are increased equally in both conditions. Clonus is more frequent in diffuse sclerosis.

Speech Disturbances.—These are more marked in diffuse sclerosis (80 per cent.). The speech may be either scanning, dragging or indistinct. Aphasia may also occur in diffuse sclerosis, but has not occurred in pseudo-sclerosis.

Affections of Other Nerves.—The cranial nerves are more frequently affected in diffuse sclerosis than they are in pseudo-sclerosis. In the former optic neuritis, optic atrophy, optic pallor, amblyopia with normal fundus, hemianopsia, transitory diplopia, and strabismus have each been noted once; nystagmus and abducens paralysis, three times. Pupillary disturbances, i. e., either inequality, slow reaction or no reaction, may be present in diffuse sclerosis, but in pseudo-sclerosis the pupils react normally. In 11 cases of diffuse sclerosis facial paralysis occurred; this was never seen in pseudo-sclerosis. Hypesthesia and hypalgesia were present in 9 cases of diffuse sclerosis, but in only 1 of pseudo-sclerosis. Forced movements and tic occurred in a number of cases of diffuse sclerosis, but in only 3 of pseudo-sclerosis. Trophic and vasomotor disturbances and muscular atrophy are much more common in diffuse sclerosis. This is also true of bladder and rectal disturbances. Remissions occur in pseudo-sclerosis, but do not in diffuse sclerosis. The duration of the two affections is about equal.

It will be noted that the symptoms of these conditions are very similar, and it seems possible that, clinically as well as pathologically, as will be shown further on, this elaborate attempt to distinguish between the two diseases may be to make a distinction without much of a difference. As has been said, some authorities seem to be tending to this view.

Thus A. Strümpell,[11] in reporting 2 cases of pseudo-sclerosis, mentions that out of 4 cases of this disorder reported by him a peculiar anomaly which seemed to connect this disease with diffuse sclerosis had existed twice, i. e., the white matter was anemic and hard. This was also noted by Westphal in one of his cases. Strümpell concludes as follows: We have to do with the picture of a disease which in its details and in its chronic course resembles markedly multiple sclerosis, but its pathologic basis has never been found in any case. In most of the sections thus far made, nothing has been found. . . . In some of these cases a peculiar dense, leathery consistency of some parts of the brain substance has been observed. No histologic cause for this is known. Such cases seem to act as transitional cases between pseudo-sclerosis and those in which the entire cerebrum possesses this dense, tough consistency, and which have been spoken of as instances of diffuse sclerosis.

Oppenheim[12] remarks that in many of its symptoms diffuse sclerosis resembles pseudo-sclerosis, in which anatomically there is an increased consistence of more or less of the tissues, so that even anatomically it is difficult to draw a distinct line between pseudo-sclerosis and diffuse sclerosis. He concludes as follows: There is a certain form of disease resembling multiple sclerosis which occurs especially in childhood, although it may appear later. It is characterized by early dementia, a special variety of tremor and disturbance of speech, muscle tremor, the absence of nystagmus, of affection of the optic nerve and of tendon phenomena. This condition is known as pseudo-sclerosis. Anatomically it is either absolutely negative or there may be a slight degree of diffuse cerebral sclerosis. The same symptom-complex, or one approaching more closely to dementia paralytica, may be associated with a true diffuse cerebral sclerosis and degeneration of the pyramidal tracts. This condition can not be diagnosed during life. The combination, occurring in childhood, of progressive spastic paralysis with progressive dementia may arouse the suspicion that such a diffuse sclerosis exists. On account of the rarity of multiple sclerosis in early childhood it is proper to make, in such patients, a probable diagnosis of diffuse sclerosis, but in such cases the question must always be left open, whether pseudo-sclerosis or diffuse sclerosis exists. These conditions must be differentiated from multiple sclerosis and paretic dementia. Weiss says that the absence of nystagmus, changes in the eye-grounds, sensory disturbances and localized muscular atrophy distinguish diffuse sclerosis from multiple sclerosis; but a reference to Frankl-Hochwart's table will show that all of these symptoms, with the exception of the last, may be present in diffuse sclerosis, though rarely.

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[11] Strümpell: Deut. Zeit. f. Nervenheilk, 1898, xii, p. 115, and vol. xiv, 1899, p. 348.

[12] Nervous Diseases, 4th Edition, p. 359.

It is evident from the foregoing that so far as our present knowledge goes the diagnosis of this condition during life is exceedingly doubtful. The peculiar features of the case reported are the advanced age of 53 years when the symptoms began, the absence until late of marked mental changes, the absence of marked speech disturbances, and the presence of the Argyll-Robertson pupil. His symptoms differed from those usually found in multiple sclerosis, in the absence of nystagmus, great increase of the tendon reflexes, and scanning speech. It may be worthy of note that until four years previous to the onset of the symptoms the patient had used alcohol freely.

PATHOLOGIC REPORT AND REMARKS BY DR. SPILLER.

Sharp distinctions between the pseudo-sclerosis and the diffuse sclerosis are impossible either by means of the symptoms or the pathologic findings. It is true that Strümpell and Frankl-Hochwart made the diagnosis of pseudo-sclerosis each in one case during the life of the patient; Strümpell did not form the correct diagnosis until he found he had erred in a case with similar symptoms that came to necropsy, and Frankl-Hochwart at first believed that his case was one of multiple sclerosis. It is not surprising, therefore, that when the patient described by Dr. Potts was seen soon after his admission to the hospital in 1900 by Dr. C. K. Mills and myself, he was supposed by us to have multiple sclerosis.

In pseudo-sclerosis the findings should be negative, and in the diffuse sclerosis a more or less intense proliferation of neuroglia throughout the brain and spinal cord should be present, but transitional forms between the two disorders have been established by the pathologic findings. The neurogliar proliferation, however, does not form the distinct areas of degeneration seen in multiple sclerosis.

I have found the reports of 14 cases of pseudo-sclerosis with necropsy, and yet the findings were not negative in all of these. In one of Westphal's cases the brain was very firm (Case 1), as it was also in one of Strümpell's cases (Case 2), and in one of Langer's cases (Case 1). Frankl-Hochwart regards such cases and the case of Burk as transitional forms. In Bäumlin's case with necropsy there were slight spinal meningitis and some loss of nerve fibers in the spinal cord. In a number of cases of pseudo-sclerosis the brain was not examined microscopically. In two of Strümpell's cases the crossed pyramidal tracts appeared to be slightly degenerated (Cases 1 and 2). In Frankl-Hochwart's case the frontal lobe was atrophic. In one of Fickler's cases the cerebral gyri were diminutive, the pons and medulla oblongata were very small, the crossed pyramidal tracts were less deeply stained and contained more neuroglia tissue than was found elsewhere in the white matter. In Fickler's other case the pons and medulla oblongata were unusually small and hard.

Strümpell, in reporting the negative findings of his third case, remarks that pathologic changes in the central nervous system must have existed, but they were not of so gross a character that they could be detected by the usual histologic methods. Schultze found thickened vessels and some swollen axis cylinders.

It may be well to give the findings in the cases of pseudo-sclerosis with necropsy. With one exception (Babinski) I have obtained the original reports of these cases and have abstracted the findings.

1. In C. Westphal's[1] first case the gyri were small, especially in the temporal and occipital lobes, the white matter of the brain was anemic and very hard, especially in the posterior portions. The microscopic examination of this case was negative.

2. Westphal's Case 2. Microscopically and macroscopically the central nervous system was normal.

Fickler includes the second case of Langer[13] as one of pseudo-sclerosis, but Strümpell thinks both cases probably belong to this group.

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[13] Wiener Med. Presse, vol. xxv, 1884, p. 698.

3. In Langer's first case the brain substance was hard, but no microscopic changes were found in the central nervous system. The patient had had convulsions and had been unable to swallow, and Langer says that his condition resembled hydrophobia when he attempted to swallow.

4. In his second case the symptoms and pathologic study are very briefly reported. No changes were found in the central nervous system.

5. Babinski.[14] The cerebral pia was a little edematous. The brain and spinal cord appeared to be normal. Microscopically the tissue was normal, except that there was some slight sclerosis in certain places. The brain was not examined microscopically.

6. Francotte.[15] Macroscopically and microscopically nothing abnormal was seen. The brain and cord were not examined by the Marchi method.

7. In Strümpell's[11] first case the consistency of the brain was normal; sections from the spinal cord stained by the Marchi method showed a moderate amount of black dots scattered diffusely, but perhaps a little more numerously in the crossed pyramidal tracts. The crossed pyramidal tracts in the cervical region were less deeply stained, and there seemed to be a slight loss of nerve fibers in these areas.

8. In his second case the brain was remarkably hard and was almost of the consistency of leather in the occipital lobe. The lateral ventricles were dilated. The crossed pyramidal tracts seemed slightly degenerated in the upper part of the cord. The tissue from the brain, although it had felt so hard, appeared to be normal under the microscope. The Marchi method probably was not employed in this case.

Strümpell regards the cases with hardening of the central nervous system as transitional forms between pseudo-sclerosis and diffuse sclerosis, and he thinks syphilis may be an important cause of both processes.

9. In Strümpell's third case the central nervous system was normal macroscopically and microscopically. Marchi's method was not employed.

10. F. Shultze[16] says, in the brief report of his case, that he found thickened vessels and some swollen axis cylinders. The cerebral cortex was not examined microscopically.

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[14] "Etude anatomique et clinique sur la sclérose en plaques," Paris, 1885.

[15] Annales de la Soc. Médico-Chirurgicale de Liege, vol. xxvi, 1887, p. 308.

[16] Lehrbuch der Nervenkrankheiten, vol. i, 1898, p. 285.

11. Bäumlin[17] found chronic leptomeningitis of the brain and some spinal meningitis, slight loss of nerve fibers in the cord, congestion of the brain, and slight dilatation of the lateral ventricles. The brain was not examined microscopically. Marchi's method was not used.

12. Frankl-Hochwart[10] found the Pacchionian granulations very numerous in his case. The frontal lobe was atrophic, but otherwise there were no macroscopic or microscopic alterations.

13. Fickler[18] (Case 1) observed sclerosis of basal vessels, and the fissures of the cerebrum were deep and the gyri small, the pons and medulla oblongata were very small and hard, and the pyramidal tracts were slightly degenerated. The brain and spinal cord were examined microscopically, but not by the method of Marchi.

14. Fickler (Case 2.) The pons and medulla oblongata were small and hard. Microscopic examination does not appear to have been made.

We may compare the findings in these cases of pseudo-sclerosis with those obtained by Hugo Weiss[9] in a case of diffuse sclerosis. The central nervous system was very hard. The brain on microscopic examination appeared to be normal. The neuroglia was proliferated in the spinal cord, and numerous spider cells were found. The septa were two or three times the usual thickness. The walls of the vessels, even of the smaller vessels, were thickened, and perivascular cellular infiltration was present. The nerve cells were normal.

The findings in other cases of diffuse sclerosis as given by Weiss have been: Distinct proliferation of the neuroglia, greater at some parts of the central nervous system than at others; perivascular round-cell infiltration, many spider cells, thickened and numerically increased vessels, and chronic meningitis. Degeneration of distinct systems of fibers does not occur.

It is evident, therefore, that sharp distinctions between the findings of pseudo-sclerosis and those of diffuse sclerosis can not be made, and that the differences are probably chiefly in the degree of the alteration and not in the character of the alteration. The unusual firmness described in some of the cases of pseudo-sclerosis must be caused by a proliferation of the neuroglia, even though this proliferation can not be detected by the microscope.

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[17] Deutsche Zeitschrift für Nervenheilkunde, vol. xx, 1901, p. 313.

[18] Deutsche Med. Wochenschrift, No. 11, Dec. 15, 1905, p. 1886.

NECROPSY.

The case that Dr. Potts and I report may be regarded as one of pseudo-sclerosis, or at least as a transitional form. It is probably the first case that has been observed in a colored person. The necropsy was made by Dr. F. P. Gay, July 29, 1902. The pathologic diagnosis was hardening of the brain and spinal cord; chronic diffuse nephritis; gummata of liver; acute serous pericarditis; acute fibrinous pleurisy.

The brain was extremely hard in all parts and the blood vessels throughout were very prominent. On section, the brain was markedly firm in both the gray and white matter. There was no evidence of gross pathologic lesions. The white substance was more firm than the gray. The cortex was slightly increased in thickness and was homogeneous and extremely firm in consistence. The injection of the capillary vessels was extremely marked throughout the brain. The basilar artery was rigid.

The spinal cord was extremely firm and showed no areas of degeneration. The entire brain, when removed from the cadaver, felt as though it had been partially hardened by some fluid.

The microscopic examination was made by me. The cerebral pia was slightly thickened and showed an unimportant round-cell infiltration. Sections from the para-central lobule stained by the Weigert hematoxylin or acid-fuchsin method presented no distinct signs of degeneration; but some of the small capillaries within this part of the brain were thickened and some were calcified. The cells of Betz, stained by thionin, were normal, except that occasionally slight diffuse staining was detected. Slight round-cell infiltration was seen about some of the small vessels within the paracentral lobule, but this was not important. In some parts of the paracentral lobule hemorrhages within the perivascular spaces were found.

The medulla oblongata appeared to be normal, both by the Weigert hematoxylin and the acid-fuchsin stains.

The neuroglia of the white matter of the spinal cord was possibly slightly increased in amount, and occasionally a somewhat swollen axis cylinder was found. The nerve cells of the anterior horns stained by thionin appeared to be normal in the cervical and lumbar regions. No alteration could be detected in the spinal cord by the

Weigert hematoxylin method, and there was no round-cell infiltration within the spinal cord or spinal pia. The Marchi sections of the cord showed a very few black dots in the crossed pyramidal tracts, so few that they are of questionable importance, and were not unlike what Strümpell found in one case.

Note.—Since this paper was prepared a case of diffuse sclerosis has been reported by Rebizzi.[19] He expresses opinions regarding the relation of the pseudosclerosis to the diffuse sclerosis similar to those set forth in this paper.

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[19] Rebizzi: Rivista di patologia nervosa e mentale, Feb., 1905, p. 57, and March, 1905, p. 105. Abstract in Review of Neurology and Psychiatry, June, 1905, p. 420.

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CHRONIC ANTERIOR POLIOMYELITIS, WITH THE REPORT OF A CASE WITH NECROPSY.

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CLINICAL NOTES AND REMARKS BY DR. MOLEEN.

Owing to the diversity of opinion with reference to the classification of spinal-cord lesions attended with muscular wasting, with or without spasticity, and with or without the presence of the deep or tendon reflexes, the assigning of the following case presents a somewhat difficult problem, and for this reason it is deemed wise to first detail the clinical notes of the case before discussing those points, which, to the author, were obstacles in arriving at an appropriate clinical diagnosis.

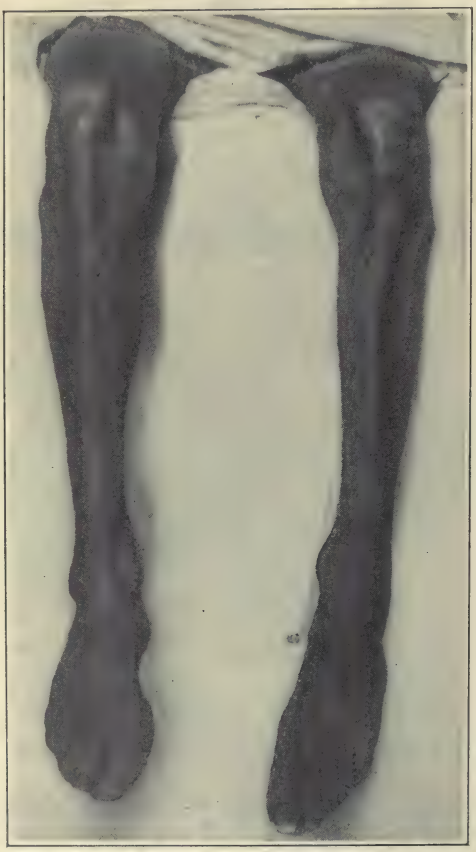
J. Ten W., aged thirty-seven years, laborer, though he did mostly upholstering work, was first seen October 3, 1903, at which time the following notes were made: He was born in St. Clair, Minn., but has been in Colorado six or seven years, and did not come for his health.

Family History. He denies all diseases of childhood, except measles, which he had at the age of ten years. He was perfectly well until 1890, when he had an attack of la grippe, and he had another in Colorado three years ago. His calloused hands bear him out in the statement that he has done hard work, and he says that he has been exposed in the mountains to rough weather until a few years ago, though he denies rheumatism or pneumonia. He admits having contracted gonorrhœa, but firmly denies syphilitic infection. It may be stated, however, that a physician related to him informs me that he treated him for syphilis twenty years ago.

Present History. About two weeks ago, without chill, pain, cold, fever, numbness, or the slightest symptom of an acute character, he noticed upon arising in the morning that his left foot felt heavy and that he was "limping."

He insists that he was perfectly well at the time and that the weakness was his only cause for complaint. He is certain that he is no worse to-day than when he first noticed his trouble. He has no difficulty in passing his urine nor in emptying his bowel.

FIG. 1



Showing the wasting of the lower limbs and foot-drop.

Examination, October 6, 1903, gave the following results:

He is a short—five feet, three inches—well-developed man, though he shows his age and hard work. His weight is 120 pounds.

In walking the left foot is raised considerably higher than the right, and with a jerky exaggerated flexion of the knee the foot is planted squarely on the floor with a tendency to strike the toe first.

Standing with the eyes open and the feet together, there is no perceptible swaying, which is but slight when the eyes are closed.

In the sitting posture, with lower limbs hanging, the "foot-drop" is noticeable on the left side.

There is no ataxia in the arm nor trunk muscles.

The calf of the left leg is somewhat atrophied and the muscles flaccid, though especially noticeable is the wasting of the muscles supplied by the peroneal nerve. The dorsum of the left foot, from the prominence of its tendons, seems more wasted than the right.

The circumference of the largest part of the calf: right, $12\frac{3}{4}$ inches; left, $11\frac{1}{2}$ inches. Of the thigh: right, $17\frac{1}{2}$ inches; left, $15\frac{3}{4}$ inches.

Electrical reactions: Galvanic battery is out of order and reactions cannot be taken. Faradic irritability is greatly diminished in the lower part of the left leg, otherwise it is normal throughout this limb. With the slowly interrupted current no contraction of the muscles supplied by the peroneal nerve can be obtained, even with the strongest current.

Reflexes: Knee-jerks, right is increased, especially on reinforcement. Left is slightly more increased than the right, but the knee-jerks were not increased to the degree commonly observed in spastic paralysis. Ankle clonus is absent. Tendo Achillis reflex: Right is present and about normal. Left is absent.

Deep reflexes of the forearm, flexors and extensors: Right are present. Left also and slightly increased. Biceps, triceps, and deltoid, same. Dynamometer (Tiemann, inner scale): Right = 280; left = 280. The man is right-handed. Masseter reflex is absent.

Superficial reflexes: Plantar: right is present; left is very slight. There is no evidence of the Babinski phenomenon. Cremasteric reflex: Right is present; left also is present, though not so marked as right. Lower abdominal reflex: Right is slight; left is very slight. Epigastric reflex is present.

Sensory phenomena are normal throughout the body (tactile pain, pressure, temperature, and postural senses taken).

Special senses: Eyes, all external ocular motion is normal and without nystagmus. Pupils are equal and respond actively to light and accommodation. Fields of vision are apparently normal. Fundi: There are no retinal changes, and the disk is well outlined in either eye. Hearing (watch): Right = 6–24; left = 8–24. The tuning-fork is heard best in the closed ear, and aerial conduction is greater than that of bone. Taste and smell are seemingly not impaired. The tongue is protruded in the median line, without tremor, and there is no difficulty in speech.

A diagnosis of anterior poliomyelitis was made at this time and he was under observation until January, 1904. During this time there was no apparent change in his condition, and it may be said that, though repeatedly examined, there was no wasting to be noticed in the hand nor in any other part of the body, nor was fibrillation to be seen at any time, these symptoms having been expected because of the absence of improvement.

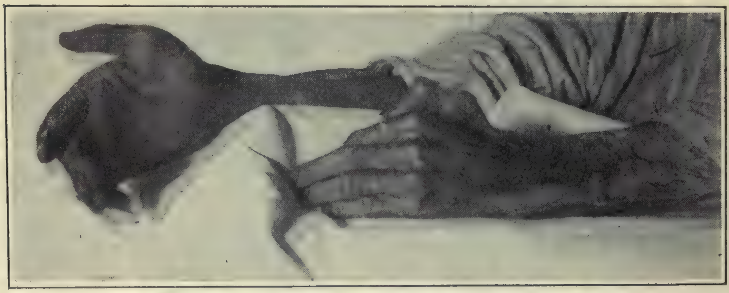
He was not seen again until January 21, 1905, when the following notes were taken:

The face is apparently drawn, probably due to emaciation, and moderate emotion (laughter) is not evidenced in the facial expression. All voluntary motion of the facial muscles, though weak, is equal on the two sides. The tongue is protruded about half an inch beyond the incisor teeth, shows a decided fibrillary tremor throughout, and is markedly furrowed from wasting. It is protruded in the median line.

He responds correctly to questions asked him, though with very slow enunciation, drolling all words requiring the use of the pharyngeal and labial muscles, but more particularly the lingual.

He states that six weeks ago he "had his palate cut" (referring to the uvula), from which time he noticed difficulty in swallowing and speaking.

FIG. 2



Showing the wasting of the hands and forearms.

There is to be noticed in the most prominent muscles—*e. g.*, biceps, deltoid, pectoral, gluteal, etc., linear depressions parallel to the muscle fibres, apparently wasted strands, between which lines of twitching are to be seen. This twitching is also to be plainly seen in the spinal muscles. Tapping the muscles increased the frequency though not the degree of the twitching.

The greatest wasting is to be observed in the hand muscles on both sides, more particularly the adductors of the thumb. The thenar and hypothenar eminences are completely wasted, and on extending the hands a decided, somewhat coarse tremor of the right thumb is noticeable (Fig. 2).

The left lower extremity is smaller in size than the right, and the group of muscles innervated by the left peroneal nerve is entirely wanting.

Plantar flexion of the left foot is somewhat weaker than of the right, while no dorsoflexion of the left foot can be accomplished.

The muscular strength in the shoulder, arms, and forearms is fairly well preserved and equal. Dynamometer, right = 110; left, = 100.

The man walks raising the left knee markedly higher than the right, and the left foot is swung farther forward and placed flatly on the floor and turned slightly outward. His gait is little if at all altered upon closing the eyes. He is able to stand with the feet together and eyes open, though he sways slightly, and the swaying is somewhat increased when the eyes are closed. The increased effort in the latter instance seemingly excites a fine tremor of the face and neck muscles.

The arm and trunk muscles are free from ataxia.

Measurements of the extremities: Arms, 4 inches above the olecranon; right = 8¾ inches; left = 8¼ inches. Arms, 4 inches below the olecranon; right = 8 inches; left = 7½ inches. Legs, 4 inches below tibial tuberosity; right = 10¾ inches; left = 9½ inches. Thighs, 8 inches below anterior superior spine, right = 15 inches; left = 12½ inches.

Electrical reactions: galvanic (chloride of silver fifty-cell battery was used, sponges moistened in alkaline tepid water, and interruptions made with the handle; current strength in milliampères):

Tibialis anticus: right, KaClC 10, KaOC 10, AnClC none, AnOC none; left, no response to the strongest current; peronei same.

Tibialis posticus: right, KaClC 6, KaOC 12, AnClC 10, AnOC none; left, KaClC 2, KaOC 3, AnClC 5, AnOC 20.

Flexor carpi radialis: right, KaClC 2, KaOC 15, AnClC 6, AnOC 25; left, KaClC 2, KaOC 20, AnClC 3, AnOC 15.

Extensor communis digitorum: right, KaClC 4, AnClC 3; left, KaClC 4, AnClC 2 (weaken rapidly).

Both the right and left deltoid muscles give anodal and cathodal contractions at 3 milliampères.

Masseters are equal: KaClC 1½, AnClC 3½. Sternocleidomastoid muscles are equal: KaClC 2, AnClC 3.

There can be no responses obtained from the thumb muscles of either hand with the strongest currents.

Faradic: (No. 8 chloride of silver battery, No. 2 secondary coil, 26 wire; two cells in circuit and current strength indicated in fractions of the rheostat circumference, unless otherwise indicated).

Tibialis anticus: Negative, right, ⅓; left, none with full current. Positive, right, ½; left, none with full current.

Rapid interruptions the same.

Tibialis posticus: Right, ½; left, ½ (negative).

Extensors of both legs respond equally to ½; flexors, ¾.

Flexor communis digitorum: left, ⅓; right, same.

The muscles of both arms and forearms show an increased irritability. Facial muscles are about normal, electrically.

Reflexes: Knee-jerks, right, increased; left, also. The effect is little changed by reinforcement. A slight ankle clonus of short duration is present in the right foot, which is very easily exhausted. Left absent. Deep reflexes of the forearm: Flexors and extensors present, those of the left side are seemingly more pronounced than those of the right side. Biceps, triceps, deltoid, and pectoral reflexes are present. Masseter reflex is absent.

Superficial reflexes: Plantar, right is present; left is absent. There is no evidence of the Babinski phenomenon, and the dorsal foot reflex lately described by Mendel[1] is negative. Cremasteric reflex: right is slight; left is absent. Lower abdominal reflex apparently is absent. Epigastric reflex is slight.

All sensory phenomena (tactile, temperature, pain, pressure, and postural) are normal and equal.

Owing to lack of control of the tongue, it is difficult to accurately test the sense of taste, though it is seemingly normal, as is also the sense of smell.

Eyes: Pupils are equal and respond equally to light and accommodation. Ocular motion is equal in all directions. The fields of vision (taken with the hand) are evidently not contracted. The disks, while pale, show no degeneration and the vessels are not changed.

Hearing is about as previously recorded.

The foregoing notes were taken during several examinations made between January 21 and 29, 1905, to which the following general observations may be added: Inability to control emotion; the extremes only were evident in the facial expression; slight laughter was often followed by tears without provocation; increasing dysphagia and dysarthria; increasing difficulty in coughing and expectorating apparently due to lack of control of the respiratory, pharyngeal, and lingual muscles.

When seen January 29, 1905, at 1 P.M., the temperature was 99.5° and pulse 100, and he was sleeping quietly. Refusing nourishment, he slept the greater part of the afternoon, death taking place at 7 P.M. of the same day, the increase in pulse and respiration rate and volume being the only change noticed, the attendants are certain, however, that no marked changes in respiration (*e. g.*, laryngeal spasm, Cheyne-Stokes breathing) were to be noticed.

Autopsy was made three hours after death. The examination was limited to the spinal cord and brain. The body was quite warm. The spinal cord and brain were removed, the cerebellum, brain stem and cord being retained for examination. The membranes were not adherent, no vascular change was noticed, and the ventricles were apparently normal. No gross changes were observed in the cerebral hemispheres. The retained portions were submitted to Dr. William G. Spiller for examination.

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[1] K. Mendel. Neurologisches Centralblatt, 1904, v. xxiii, No. 5.

SUMMARY. A man, aged thirty-seven years, probably syphilitic, is afflicted with sudden paralysis of the group of muscles innervated by the left peroneal nerve; complete loss of faradic irritability followed in a week, and this was followed by rapid wasting and slight diminution in size of the entire leg; the deep reflexes were present and increased, but not excessively, and in that spinal segment represented by the absolute atrophy they were lost; the symptoms were confined to the left lower extremity and were without change during three months; no fibrillary tremor, sensory disturbance, hand weaknesses, nor cranial nerve symptoms were present. One year later there were slight general emaciation, complete wasting of the thenar and hypothenar eminences, and, to a less extent, of the interossei muscles; atrophy and paresis of the tongue and pharynx, fibrillation throughout the body, including the tongue, and tendon reflexes still moderately increased. There were no marked electrical changes, except in absolutely wasted muscles, and no bladder, bowel, nor sensory disturbances. He was becoming more emotional, dysphagic and without dyspnœa, though he had a rapidly increasing pulse and respiration. Termination was death.

When first observed the recording of the case as acute anterior poliomyelitis, in view of the rapid onset and the absence of evidence of involvement of other portions of the cord, was justifiable. The only points apparently opposed being the age of the patient and the absence of prodromes or febrile disturbance.

The fact that no evidences of extension nor involvement elsewhere could be noticed after three months seemingly supported this diagnosis.

When, however, wasting of the hands, fibrillation, dysarthria, and dysphagia were added after the lapse of a year, one would be justly suspicious of (*a*) progressive spinal muscular atrophy (especially a peroneal type), (*b*) amyotrophic lateral sclerosis, or (*c*) chronic poliomyelitis—three conditions which have been considered by some authorities as synonymous—the extremely wide variations in individual cases notwithstanding.

It seems pretty well established that in pure cases of progressive spinal muscular atrophy the wasting invariably precedes the paralysis, and certainly the great majority of cases first manifest weakness in the hands (nine-tenths, according to Gowers). Conspicuous also is the advent of fibrillary twitching in the gradually weakening muscles, as well as in those about to be attacked, in which the weakness had not been noticed, and in any event before paralysis had supervened.

The electrical reactions in these cases of progressive spinal muscular atrophy persist until the wasting is intense.

Clinically, perhaps, this case more closely conforms to the type known as "peroneal," which since the thesis of Dr. Tooth and the observations of Dr. Hoffmann, has, in the opinion of most authorities, merited a separate classification, and some have referred to it as "neuritic" (perhaps more properly "neurotic," from the "Progressive Neurotische Muskelatrophie" of Hoffmann), in view of its occurrence in several members of the same family, in the majority of instances. That it occurs, as a rule, in the second half of childhood—*i. e.*, before the twentieth year of life, seems to be generally accepted, though Oppenheim thinks the commencement may be as late as the fourth decade.

Some observers have used the term "neuritic" advisedly, because of the belief that the peripheral nerves are primarily affected. This is questioned by Dana, who believes the cord to be first attacked, and quotes Marinesco as having recently confirmed this view, mentioning a citation by the same author of a case beginning as a "typical acute anterior poliomyelitis."

As rather against than for the placing of this case under this heading, the following features deserve mention: The occurrence of the disease earlier in life as a rule; usually hereditary involvement; frequent sensory disturbances (pain, numbness, etc.); usually abolition of the tendon reflexes in the extremity affected; slow development; atrophy usually extreme; lastly, conspicuous by its absence in the cases reported is the mention of bulbar symptoms.

As tersely expressed by Dana, the hereditary or "leg type" of progressive muscular atrophy is characterized by its beginning in the legs, by much sensory disturbance, typical degeneration reactions and hereditary or family history.

Is this a case of amyotrophic lateral sclerosis?

It is well before considering this condition to disregard the question of synonymy, and with Oppenheim, Dana, Beevor, and others, return to the original distinctive classification of Charcot. This disease occurs most frequently after thirty, women being more often affected than men; in true types the hands are first affected, and the spasticity is marked; the first symptom is usually rigidity; clonuses, including a masseter clonus, are frequent; flaccid paralysis never inaugurates the disease, and the onset is nearly always gradual. Oppenheim only records an acute beginning; as a rule, each muscle wastes very gradually.

In the study particularly of the clinical side of the case, however strong our convictions may lead us to the heading of progressive muscular atrophy, the consideration of the distinctive points of the third of the "trio," chronic anterior poliomyelitis cannot be overlooked. This line of distinction, one must admit, in the light of our present knowledge, is as yet fine.

While partial recovery is the rule, though more especially in the acute form of anterior poliomyelitis, yet cases do become subacute or chronic, and extension or probably the independent involvement of other segments, including the bulb, are then not uncommon, and in fact bulbar paralysis very frequently closes the scene in all three of the conditions mentioned—viz., progressive spinal muscular atrophy, amyotrophic lateral sclerosis, and chronic anterior poliomyelitis.

In chronic anterior poliomyelitis the paralysis precedes the wasting, the palsy is absolute, flaccid, without rigidity, and involves groups of muscles physiologically related, while in the other affections previously considered the muscles concerned are anatomically related. Syphilis is a frequent cause in advanced life of the chronic cases; all electrical irritability is rapidly lost in the group of muscles affected, and neighboring muscles are altered in proportion to the degree of subacute extension, this alteration becoming less marked farthest from the cord representation of the principal group involved. A predilection of the peroneal group in all cases of anterior poliomyelitis is notable.

The mention of fibrillary twitching has been purposely omitted heretofore, for the reason that it is seemingly common to all chronic muscular atrophies, and Gowers, in his broad view of the three diseases as one, states that it is not invariable, nor is it confined to this disease; and Oppenheim has to say: "Progressive muscular atrophy can never be diagnosed from fibrillary tremor, as this symptom may occur in healthy persons after excesses, in emaciated individuals when exposed to cold, and in neurasthenic and hypochondriac patients."

While disposed to rest the diagnosis here, I am constrained to admit that against this disposition is the presence or increased activity of the deep reflexes which in the cases previously reported as such was absent; the explanation offered with reference to the electrical alterations in the segments immediately adjacent to the actual lesion, it is reasonable to suppose, will hold good here, and possibly establish a point of distinction with future observation.

It is therefore evident that while no resting place is absolutely fitted for the reception of this particular case, the wide latitude of which chronic anterior poliomyelitis admits, and the fact that here are found fewer opposing factors than in the other mentioned conditions render its placing here at least permissible.

The emotional symptoms at the close of the case are, as noted by Dana, common evidences of bulbar lesions, either primary or secondary.

PATHOLOGICAL NOTES AND REMARKS BY DR. SPILLER.

The case which Dr. Moleen has studied clinically so carefully is worthy of the attention he has given to it. I know of no case with necropsy like it in American or English literature, and it should, therefore, be of value in the study of the muscular atrophies. This is a subject that is still causing much difficulty in classification. The commencement of the symptoms in Dr. Moleen's patient with peroneal palsy, resembling the neurotic muscular atrophy, is characteristic of certain lesions of the cells of the anterior horns. Baumann,[1] in a clinical study of 85 cases of acute anterior poliomyelitis, has shown that the muscular supply of the peroneal nerve is especially liable to become paralyzed in this disease.

Dejerine[2] reviews the opinions held regarding motor localization within the spinal cord. According to him, it is not segmental (hand, forearm, arm), as Van Gehuchten and De Buck believed; it is not diffuse (Marinesco), each muscle is not separately represented (Sano), but the localization is of the root type. An examination of the diagram given by Dejerine, p. 780, shows that the peroneal supply is represented in the fourth and fifth lumbar and first sacral segments. We can, therefore, understand why, in Dr. Moleen's patient, the paralysis began in the peroneal group.

My report is as follows:

Only the spinal cord and brain stem of this case were sent to me for examination.

Lumbar Region. The white matter by the Weigert hæmatoxylin stain does not appear to be degenerated. The small vessels of the anterior horns are much congested and are, therefore, very prominent by this stain. Those of the posterior horns are also congested, but not so much so as are the vessels of the anterior horns. Small hemorrhages are numerous in the gray matter, especially in the anterior horns, but are also present in the posterior horns. The posterior roots are well stained. The anterior roots of the lumbar and sacral regions show in places distinct degeneration, but these roots are far better preserved than are the cells of the anterior horns. There is no round-cell infiltration within the spinal cord or pia. The nerve cells of the anterior horns are much diminished in number, but those that still persist appear to be almost normal, and only occasionally one is seen showing chromatolysis, displacement of the nucleus, or much pigmentation. A slight recent degeneration in the crossed pyramidal tracts is seen *only* by the Marchi method.

Middle Thoracic Region. Small hemorrhages are numerous in the gray matter, in the posterior as well as in the anterior horns, and are found, but much less numerously, in the white matter near the gray. The Marchi method reveals a very slight degeneration of the crossed pyramidal tracts and a few scattered black dots elsewhere in the anterolateral columns.

First Thoracic and Eighth Cervical Segments. The small hemorrhages are very numerous in the anterior and posterior horns, and are found also in the white matter near the gray. In this portion of the cord the nerve cells of the anterior horns have almost disappeared. There is hemorrhage also within the pia, but nowhere is there any round-cell infiltration.

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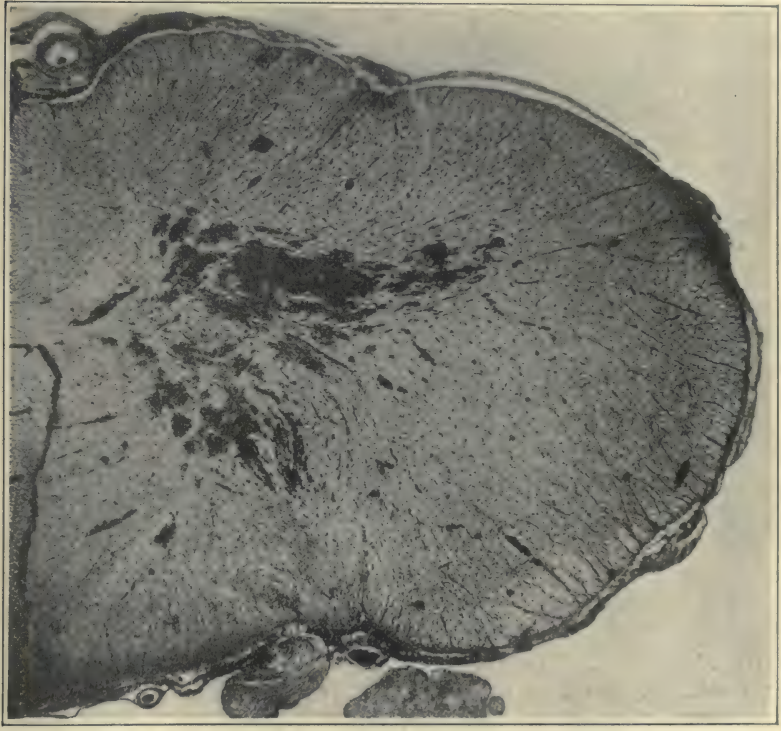
[1] Monatsschrift f. Psychiatrie und Neurologie, June, 1905, p. 485.

[2] Sémiologie du système nerveux, p. 789.

Medulla Oblongata. Small hemorrhages are numerous here also. Intramedullary fibres of the twelfth nerve have disappeared by degeneration. Unfortunately the nuclei of the twelfth nerves have been injured at the time of the necropsy, but a marked disappearance of nerve cells in these nuclei may be determined. The nucleus ambiguus contains few cells. The intramedullary fibres of the ninth and tenth nerves are much atrophied. A very slight degeneration of the anterior pyramids is seen only by the Marchi method and is less than the degeneration of the crossed pyramidal tracts of the spinal cord.

FIG. 3



Showing the numerous hemorrhages within the gray matter of the spinal cord.

There are few cells in the nuclei of the seventh nerves and the intramedullary fibres of these nerves are much degenerated. Small hemorrhages are not so numerous in the pons. The cells still persisting in the facial nuclei stain fairly well, except that here and there a cell may be found distinctly degenerated.

A few small hemorrhages are found in the cerebral peduncles.

The important alterations in this case are disappearance of many of the nerve cells of the anterior horns of the spinal cord and motor cranial nerves in the medulla oblongata, of the anterior roots of the lumbar and sacral regions (cervical region was injured), and of the motor nerves of the medulla oblongata, and numerous small hemorrhages in the gray matter, especially in that of the spinal cord.

It is very questionable whether we should make sharp distinctions between chronic anterior poliomyelitis and progressive spinal muscular atrophy. In the first the paralysis is supposed to develop within a few days or a few weeks, the atrophy appears later, and entire muscles or groups of muscles are paralyzed, the course of the disease is more rapid, and the paralysis develops first in the muscles of the lower limbs or shoulders. In the progressive spinal muscular atrophy the paralysis is proportionate to the atrophy, one muscle fibre after another is affected, and the course is longer than in the chronic poliomyelitis. These are distinctions that are not always regarded, and it is very evident that some of the reported cases of so-called chronic anterior poliomyelitis might just as well be labelled as examples of progressive spinal muscular atrophy.

The cases of either type with necropsy are very rare. To Oppenheim the credit of reporting the first typical case of chronic poliomyelitis with necropsy has been justly given. In Oppenheim's[1] first case the weakness began in the right arm. The vessels of the cord were not distinctly altered.

In the case that S. G. Webber[2] reported in 1875 weakness began in the left knee and soon the left leg gave out. Atrophy did not appear until after the weakness. Sensation was not affected. The nerve cells of the cervical region were much diseased and were not numerous. There was some perivascular bulbar infiltration. The white matter of the cord appeared as a homogeneous mass, not colored by carmine; occasionally an axis-cylinder could be seen. The lumbar and thoracic portions of the cord could not be satisfactorily studied. The nerve cells of the hypoglossal nuclei were few in number and many axis-cylinders were swollen. The posterior and anterior roots were also affected.

This case is important, as it was reported about thirty years ago, and clinically seems to have presented the symptoms of chronic anterior poliomyelitis, but pathologically it appears to have been a case of diffuse myelitis. The duration of the symptoms was about one year.

It is true that Nonne[3] included as cases of subacute anterior poliomyelitis those reported by Eisenlohr, and Landouzy and Dejerine, and as a case of chronic anterior poliomyelitis the case of Dreschfeld. These three cases he acknowledges were very different from one another, and he makes three types of chronic anterior poliomyelitis: 1. The circumscribed form (Eisenlohr's case), in which the lesion is confined to a limited area. 2. A form in which paralysis and atrophy *en masse* occur quickly, show a tendency to arrest or to complete restoration, and may possibly be the result of acute or chronic infection (Landouzy and Dejerine's case). 3. A form which progresses slowly and irresistibly with atrophy of nerve cells without secondary atrophy of peripheral nerves (Dreschfeld's case) or with this atrophy (Oppenheim's case).

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[1] Archiv f. Psychiatrie, 1888, Bd. xix. p. 381.

[2] Transactions of the American Neurological Association, 1875, p. 55.

[3] Deutsche Zeitschrift f. Nervenheilkunde, 1891, Bd. i. p. 136.

Oppenheim, in a later paper, does not accept as cases of chronic or subacute anterior poliomyelitis those referred to by Nonne, and it seems to me that Oppenheim is right in this opinion.

Eisenlohr's case[1] was remarkable on account of the limitation of the process. The weakness and atrophy affected only the right upper limb and persisted about five years. The nerve cells were degenerated only in the right anterior horn between the sixth cervical segment and the first thoracic. The white matter was normal. It is difficult to decide why this case was regarded as one of subacute anterior poliomyelitis and not as one of progressive spinal muscular atrophy.

Dreschfeld[2] regarded the case he reported as one of chronic anterior poliomyelitis. Weakness began in the right foot and leg. The tendon reflexes of the upper limbs were very prompt. The nerve cells of the anterior horns were much degenerated. Lateral sclerosis was found in the cervical region. It seems doubtful whether this should be regarded as a case of amyotrophic lateral sclerosis or as one of chronic anterior poliomyelitis.

Landouzy and Dejerine[3] did not regard their case as one of chronic anterior poliomyelitis.

A case that probably should be classed as one of chronic anterior poliomyelitis with necropsy was reported by Richard Ewald.[4]

The case that Nonne reports began with weakness in the left shoulder and left upper arm. Atrophy occurred at the same time as the paralysis or followed soon after. In the last months of the disease pains were felt in the upper part of the upper limbs and in the thighs. Nonne[5] found some degeneration of peripheral nerves. as well as of nerve cells, in the spinal cord, and he refers to the fact that in Oppenheim's case the hypoglossal and spinal accessory nerves and the nerves within muscles were not entirely normal. Nonne found atrophy of nerve fibres in the white matter of the cord, in the pyramidal tracts as well as elsewhere, but not as symmetrical degeneration.

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[1] Neurologisches Centralblatt, 1882, p. 409.

[2] Brain, 1886, p. 173.

[3] Revue de méd., 1882.

[4] Ein Fall von Poliomyelitis anterior chronica, Inaugural Dissertation, Marburg, 1899.

[5] Deutsche Zeitschrift f. Nervenheilkunde, 1891, Bd. i. p. 136.

In Darkschewitsch's[1] case weakness and atrophy developed gradually in the muscles of the upper limbs, neck, back, and partly in the lower limbs. The nerve cells of the anterior horns of the cervical and thoracic regions were diminished in number, and there were numerous hemorrhages which seem to have been in the gray matter of the cord. Rarefaction of the lateral and posterior columns was observed. It is difficult to determine from the brief report why this was regarded as a case of chronic poliomyelitis and not as one of progressive spinal muscular atrophy.

In Oppenheim's[2] second case of chronic anterior poliomyelitis the weakness began in the right shoulder and right upper arm, and after a year extended to the corresponding parts of the left upper limb. Atrophy soon followed the weakness. Paræsthesia was felt in the fingers in the beginning of the disease. The nerve cells of the anterior horns of the cervical and thoracic regions were much degenerated. The columns of Burdach were also degenerated.

In the case reported by A. Dutil and J. B. Charcot,[3] the symptoms began with weakness in the upper limbs. The vessels of the cord were thickened and some small recent hemorrhages were found in the cord. This is the same case that J. B. Charcot later reported alone.

In J. B. Charcot's[4] two cases of chronic anterior poliomyelitis there was some degeneration of the anterior fundamental fasciculus—*i. e.*, of the fibres near the anterior horn. These cases possibly might be regarded as examples of progressive spinal muscular atrophy.

It is difficult two understand why Dejerine's[5] two cases should be regarded as examples of chronic anterior poliomyelitis and not of progressive spinal muscular atrophy, if a sharp distinction is maintained. In one case there was a questionable sclerosis of the interior portion of the right lateral column.

Nonne,[6] in 1896, recognized as cases of chronic anterior poliomyelitis with necropsy the two cases of Oppenheim, the two cases of his own, the case of Darkschewitsch, the case of Dutil and J. B. Charcot, and the two cases of Dejerine.

In Nonne's second case the weakness began in the shoulder muscles. The nerve cells of the anterior horns were much degenerated and rarefaction of the white matter was seen in the anterior, lateral, and posterior columns. The bloodvessels were not distinctly altered. He attributed the poliomyelitis in this case to diabetes. Nonne refers to the slight changes in the white matter observed in Oppenheim's two cases, in his own two cases, in

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[1] Neurologisches Centralblatt, 1892, Bd. xi. p. 221.

[2] Archiv f. Psychiatrie, 1892, Bd. xxiv. p. 822.

[3] Le progrès médical, March 17, 1894, p. 185.

[4] Archives de médecine expérimentale, 1895, vol. vii. p. 441.

[5] Comptes-rendus heb. des séances de la Soc. de biologie, 1895, p. 188.

[6] Berliner klin. Wochenschrift, March 9, 1896, p. 207.

Darkschewitsch's case, in Dutil and Charcot's case, and in one of Dejerine's cases.

In P. Schuster's[1] atypical case of chronic anterior poliomyelitis, which was complicated by degeneration of the posterior roots and of the columns of Burdach, the patellar reflexes were much exaggerated. The explanation was supposed to be found in the slight alteration of the lateral columns.

Max Bielschowsky[2] says there are very few cases of chronic anterior poliomyelitis reported that show evidences of inflammation. In Bielschowsky's case the weakness began in the lower limbs, in the peroneal distribution, as in the case Dr. Moleen and I report. He says that contrary to the diagnosis of neural atrophy (peroneal type) were the later developments of the disease and the absence of subjective and objective disturbance of sensation. Contrary to progressive spinal muscular atrophy were the location of the atrophy, the relatively rapid development of the symptoms, and the early loss of function in the diseased muscles. Hemorrhages were found in Bielschowsky's case in the anterior horns from the lumbar to the upper cervical region. In some places the red blood corpuscles had disintegrated, blood pigment was present, and reactive processes had occurred about some of the hemorrhages. These hemorrhages were regarded as evidences of inflammation. Slight degeneration was seen in the crossed pyramidal tracts of the spinal cord as high as the pons, but was detected only by the Marchi method and was slight, exactly as I have found in the specimens studied by me. Bielschowsky observed slight perivascular round-cell infiltration and newly formed bloodvessels. The hypoglossus nuclei also were affected. This case resembles ours very closely, although the disease in Bielschowsky's case lasted nine years. The tongue was also weak and atrophied. No statement is made concerning the condition of the patellar reflex until late in the disease, when the atrophy was intense.

In Grunow's[3] case the symptoms seem to have existed about nine months. The weakness began in the right foot and peroneal palsy developed. Pain was felt in the shoulders and back. Progressive spinal muscular atrophy was excluded because the weakness began in a lower limb, because of the wide extent of the process, the implication of intercostal muscles, and the relative rapidity of development. As in our case, peroneal palsy was among the earliest signs of the disease. The pain in this case is noteworthy. Cellular infiltration was found about the arteries of the anterior fissure and about some of the vessels of the anterior horns and white matter. Necrotic foci were present in the crossed pyramidal tracts, and there were some small hemorrhages in the gray matter of the cord.

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[1] Neurologisches Centralblatt, 1897, Bd. xvi. pp 342, 392, 449.

[2] Zeitschrift f. klin. Med., 1899, Bd. xxxvii. p. 1.

[3] Deutsche Zeitschrift f. Nervenheilkunde, 1901, Bd. xx. p. 333.

The nerve cells of the anterior horns were much degenerated. Degeneration was found in the anterior and lateral columns.

Bruining[1] has observed chronic anterior poliomyelitis in father and son, and in one of the cases obtained a necropsy. In neither case were there periods of abrupt increase in the symptoms. The weakness seemed to be proportional to the atrophy. In the father the weakness and atrophy began in the right thigh and then the shoulders became implicated. In the son the atrophy began in the muscles of the neck and extended to the shoulders and upper limbs. The nerve cells of the anterior horns throughout the cord were degenerated, but no signs of inflammation were found. The capillaries of the cord were congested and in some places blood corpuscles were found in the tissues about the vessels. Bruining is not inclined to make sharp distinctions between chronic anterior poliomyelitis and progressive spinal muscular atrophy.

Rossolimo,[2] in reporting a case of chronic anterior poliomyelitis with syringomyelia, recognized that somewhat exaggerated patellar reflexes may occur in anterior poliomyelitis, and refers to the cases of Darkschewitsch and Schuster. He recognizes as cases of chronic anterior poliomyelitis only those of Oppenheim, Nonne, Darkschewitsch, Schuster (not a pure case), Bielschowsky, and R. Ewald.

Oppenheim, in speaking of chronic anterior poliomyelitis in his text-book, says exaggeration of tendon reflexes alone is not sufficient to make the case one of amyotrophic lateral sclerosis, as exaggeration of reflexes may occur in neurasthenia.

In the monograph on poliomyelitis written by Elis Lövegren,[3] the author reports a case of subacute poliomyelitis with necropsy. The weakness began in the fingers and was followed in two or three months by atrophy of the hands. The paresis and atrophy became widespread. The patellar reflexes were exaggerated. The nerve cells of the anterior horns of the spinal cord and of nuclei of motor cranial nerves were degenerated. Vessels with thickened walls were found in the spinal cord, but there was no round-cell infiltration. The white matter of the cord was said not to be involved and the pyramidal tracts to be intact. The method of Marchi was not employed. The anterior roots and peripheral nerves showed slight atrophy. A cavity in a limited area was found in one anterior horn.

Lövegren refers to two cases of subacute or chronic poliomyelitis with necropsy reported by Philippe and Cestan,[4] in which a slight sclerosis of the anterolateral columns was found; and to a case with necropsy reported by Raymond and Philippe.[5] I have not been able to obtain the paper by Philippe and Cestan.

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[1] Deutsche Zeitschrift f. Nervenheilkunde, 1901, Bd. xxvii. p. 269.

[2] Neurologisches Centralblatt, 1903, p. 388.

[3] Sonder-Abdruck a. d. Jahrbuch f. Kinderheilkunde, S. Karger, Berlin, 1905, vol. lxi.

[4] XIII. Congrès international de médecine, Paris, 1900, comptes-rendus, section de neurologie, p. 271.

[5] Soc. de neurologie, Nov. 6, 1902, Archives de neurologie, 1902, 2 série, tome xiv. p. 537.

The case of chronic poliomyelitis very briefly reported by Raymond and Philippe[1] is as follows: The disease began at the age of fifty-two years in the extremities of the lower limbs, and is said by the authors to have been the first case of the kind. The antero-external muscles of the legs first showed atrophy and the weakness progressed simultaneously with the atrophy. The poliomyelitis was strictly limited to the nerve cells of the anterior horns. The anterior roots and peripheral nerves were secondarily affected. No mention is made of the employment of the method of Marchi or of the condition of the tendon reflexes.

The case observed clinically by Dr. Moleen and studied from the pathological aspect by me may properly, I think, be regarded as one of chronic anterior poliomyelitis. The commencement in one lower limb, the paralysis preceding the atrophy, the comparatively rapid development of the symptoms, the absence of sensory disturbances, and the pathological findings justify this diagnosis. The prompt tendon reflexes are unusual, but it is probable that Bielschowsky's patient would have shown the same condition if these reflexes had been observed earlier in the disease. The patellar reflexes were exaggerated also in Lövegren's case. The degeneration of the crossed pyramidal tracts in our case was very slight and perceptible only by the Marchi method. When the Weigert method was employed no suspicion of degeneration of the pyramidal tracts could be entertained. The case, however, shows that the distinctions between amyotrophic lateral sclerosis and chronic anterior poliomyelitis are not always to be sharply made. The Marchi method does not seem to have been employed in the examinations of many of the cases of chronic anterior poliomyelitis reported.

It is noteworthy that pain was recorded as present in the cases of Nonne (Case I.) and Grunow, and that paræsthesia occurred in Oppenheim's second case.

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[1] Revue neurologique, 1902, p. 1075.

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